

Rare extragonadal teratomas in children: complete tumor excision as a reliable and essential procedure for significant survival

Clinical experience and review of the literature



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Rare extragonadal teratomas in children: complete tumor excision as a reliable and essential procedure for significant survival. Clinical experience and review of the literature

BACKGROUND: *Extragonadal teratomas are rare tumors in neonates and infants and can sometimes show unusual, distinctive features such as an unusual location, a clinical presentation sometimes acute, and a "fetiform" histotype of the lesion. We have extrapolated, from our entire experience of teratomas, 4 unusual cases, mostly operated as emergencies; 2 of them were treated just after birth. Aim of this paper is to report the clinical and pathological findings, to evaluate the surgical approach and the long-term biological behaviour in these cases, in the light of survival and current insights reported in the literature.*

MATERIAL AND METHODS: *The Authors reviewed the most significant (Tables I and II) clinical, laboratory, radiologic, and pathologic findings, surgical procedures, early and long-term results in 4 children, 1 male and 3 females (M/F ratio: 1/3), suffering from extragonadal teratomas, located in the temporo-zygomatic region of the head (Case n. 1, Fig. 1), retroperitoneal space (Case n. 2, Fig. 2), liver (Case n. 3, Figs. 3-5), kidney (Case n. 4, Fig. 6, 7), respectively. Of the 4 patients, 2 were treated neonatally (1 T. of the head, 1 retroperitoneal T.) A prenatal diagnosis had already been made in 2 of the 4 patients, between the 2nd and 3rd trimester of pregnancy. All the infants were born by scheduled caesarean section in a tertiary care hospital and were immediately referred to the N.I.C.U.s. Because of a mostly acute clinical presentation, the 4 patients were then referred to the surgical unit at different ages: 7 days, 28 days, 7 months, and 4 years respectively. The initial clinical presentation (Table II) was consistent with the site of the mass and/or its side effects. The 2 newborns (Case 1 and 2) both with a prenatally diagnosed mass located at the temporo-zygomatic region and in the abdominal cavity respectively, already displayed, at birth a mass with a tendency to further growth. The symptoms and signs described to the primary care physician by the parents of the 2 patients suffering from intra-abdominal tumours (Cases n. 3, 4) were: swelling of the epigastrium and left hypochondrium due to a progressively growing hard mass, without impairment of the general conditions in case n.3 (teratoma of the liver), while recurrent abdominal pain lasting for the 5 months was described in case n.4 (retroperitoneal teratoma), followed by the development of an evident hard mass occupying the entire abdomen. In this case the symptoms suddenly worsened, with acute pain extending to the entire abdomen, high fever (>39° C), polypnea, anemia, deterioration of the general conditions and a rapid further enlargement of the mass. Antibiotic therapy was unsuccessful. The young child underwent a radiologic investigation (Fig. 6) that showed a large calcified mass in the left retroperitoneal space, associated with pleural effusion. In all the patients except for the Case n. 3, emergency surgical management was required and, in accordance with recommended practice, the procedure was complete exeresis.*

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RESULTS (TABLE I): All the 4 patients had an uneventful postoperative course. Clinical surveillance and tests of AFP and other markers were scheduled every 6 months for the first years and annually thereafter. At the current date they are alive, disease-free and have not suffered any recurrence with a follow-up as reported in Table I, of 7 years in case n. 2; 23 years in case n. 1; 42 years in case n. 3 and 36 years in case n. 4.

CONCLUSIONS: Some extragonadal teratomas of childhood may rarely arise in the solid organs (liver, kidney), in the retroperitoneal space or the cranio-facial region, and also show unique histotype childhood characteristics ("fetiform") which distinguish them from more common cases. Being congenital tumours, prenatal diagnosis by US scan is extremely important in order to organize proper perinatal care in appropriate facilities where it is possible to define the diagnosis and carry out emergency surgery. An emergency procedure is frequently dictated both by complications related to the mass effect and by the need to define the histology of the whole mass rather than just small biopsy specimens. Some teratomas can hide more or less extensive islands of immaturity signs of malignant transformation that are clinically evident. It should be remembered that high serum levels of alpha-fetoprotein and calcification of the imaging study, that are usually pathognomonic elements for diagnosis, may be lacking in abdominal lesions. Moreover, some additional specific diagnostic problems can be faced by either the radiologist (differential diagnosis from acquired or congenital cystic lesions, identification of the primary site of origin in the liver kidney or retroperitoneal space). Or the histopathologist (exclusion of renal metastasis of a primary gonadal teratomas of a glomerular and tubular differentiation a Wilms' tumour). The prognosis is generally benign, although the AIEOP guideline pointed out that high levels of circulating markers, including AFP, in children affected by mature or immature teratomas, could indicate the presence of micro-foci of YST, marking them out as at high risk. The UKCCSG II and the SFOP indicates AFP values exceeding 10,000 ng/ml as the threshold identifying a group of patients with a severe prognosis. The treatment indicated is early, complete exeresis, followed by a careful, extensive, microscopic examination, associated, if necessary, with adjuvant chemotherapy. Finally, to improve the prognosis, close, long-term clinical, laboratory and imaging surveillance is necessary, at shorter intervals during the first 5 years after the exeresis and annually thereafter.

KEY WORDS: Extragonadal tumors, Germ cell tumor, Teratomas

Introduction

Extragonadal teratomas are rare, and can sometimes show unusual, distinctive features unlike those commonly described. These characteristic features include: 1) an unusual location, 2) a clinical, sometimes acute, presentation 3) an unusual phenotypic appearance of the mass and 4) a "fetiform" histotype of the lesion. These ele-

ments may be isolated or associated within the same lesion. The Authors have extrapolated, from their entire experience of teratomas, 4 selected cases, mostly operated as emergencies. Aim of this paper is to report the clinical and pathologic findings, to evaluate the surgical approach and the long-term biological behavior in these cases, in the light of survival and current insights reported in the literature.

TABLE I

	Cases	Age	Sex	Site	Pre-Nat. Us	Afp	Surgery Date	Procedure	Pathology	Results
1	F.M.	7 days	M	Head: temporo-zygomatic-region	Yes	H	1989	Exeresis of mas and temporal muscle	Mature triphyllic solid-cystic terat. + temporal muscle dismembered	Alive 23 Y
2	G.C.	28 days	F	Left retro-peritoneal space	Yes	H	2005	Complete exeresis	Mature triphyllic solid-cystic terat	Alive 7 Y
3	M.R.	7 m.	F	Liver, left lobe	—	H	1970	Exeresis	Mature “fetiform” solid-cystic terat.	Alive 42 Y
4	F.A.	4 Y.	F	Left kidney	—	H	1976	Exeresis of left kidney/mass	Predominantly solid triphyllic “fetiform” terat.	Alive 36 Y

Materials and Method

We reviewed the most significant (Tables I, II) clinical, laboratory, radiologic and pathologic findings, surgical procedures, and early and long-term results in 4 children, 1 male and 3 females (M/F ratio: 1/3), suffering from extragonadal teratomas, located in the temporo-zygomatic region of the head (case N° 1 - Fig. 1), retroperitoneal space (case N° 2 - Fig. 2), liver (case N° 3- Figg. 3-5), kidney (case N° 4 - Figs. 6, 7). Of the 4 patients, 2 were treated neonatally (1 T. of the head, 1 retroperitoneal T.). Among the 4 patients, a prenatal diagnosis had already been made in 2 of them between the 2nd and 3rd trimester of pregnancy. All the infants were born by scheduled caesarean section in a tertiary care hospital and were then immediately referred to the N.I.C.U.s. Because of a mostly acute clinical presentation, the 4 patients were referred to the surgical unit at different ages: 7 days, 28 days, 7 months, and 4 years, respectively. The initial clinical presentation (Table II) was consistent with the site of the mass, a sudden volumetric increase and/or complication. The 2 newborns (cases N° 1, 2), both with a prenatally diagnosed mass located at the temporo-zygomatic region and in the abdominal cavity, respectively, already displayed, at birth, a mass with a tendency to further

growth. The symptoms and signs described to the primary care physician by the parents of 2 patients suffering from intra-abdominal tumors (cases N° 3, 4) were: swelling of the epigastrium and left hypochondrium due to a progressively growing hard mass, without impairment of the general conditions in case N° 3 (teratoma of the liver); recurrent abdominal pain for 5 months was described in case N° 4 (retroperitoneal teratoma), followed by the development of an evident hard mass occupying the entire left hemiabdomen. In this case the symptoms suddenly worsened, with acute pain extending to the whole abdomen, high fever (>39 °C), polypnea, anemia, deterioration of the general conditions and a rapid further enlargement of the mass. Antibiotic therapy was unsuccessful. Radiologic investigation (Fig. 6b) showed a large calcified mass in the left retroperitoneal space, associated with pleural effusion.

Physical Examination: The general conditions were good in all patients except case N° 4, a 4 year-old female child referred for severe respiratory distress, anemia, high fever, acute abdomen and poor general conditions. The physical findings are summarized in Table II.

Laboratory: Blood tests showed moderate anemia (Hb 3,200,000, RBC 8.9) with reduced proteinemia (g 5.6%),

Table II - *Clinical and imaging findings*

Cases	Symptoms	Physical signs	Radiology	
1) 7 days M, 1989	At birth, round fixed swelling on the right Temporo-zygomatic region	Well delimited round, fixed soft tissue mass extending in the orbit	Cranial CT calcified response	Large heterogeneous mass located in front of the temporo-zygomatic wing suture, imprinting the orbit, without bone erosion
2) 28 days F, 2005	Abdominal distension	Retroperitoneal hard mass occupying the left hemiabdomen	CT, Angio MR	Well circumscribed homogeneous noncalcified mass located in front of the kidney, with normal shape but displaced downward
3) 7 months F, 1970	Gradual increasing distension of upper abdomen	Hard mass occupying epigastric-left hypochondrium region, mobile during breathing movements	Plain Hepatic Scan: Radiol-Isotope scan	Organoid calcifications inside a medium density opacity. Cold areas in the left hepatic lobe
4) 4 years F, 1976	Recurrent abdominal pain for 5 months. Evidence of a mass with high fever (39°), anemia, polypnea, acute abdomen	Poor general conditions. Left basal pleural effusion. Voluminous tender, solid mass in the left abdomen, Overcrossing the midline. Contracted abdominal muscles	Plain Abdominal Radiographs+ i.v Pyelography before hospitalization	Organoid-like calcifications inside a large-high density mass in the left abdomen overcrossing the midline involving the upper pole of the kidney, displacing and deforming the collecting system

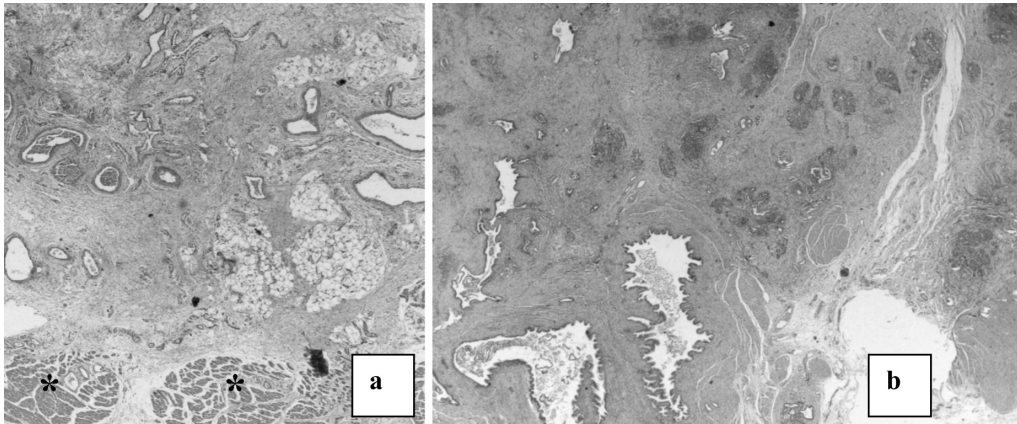


Fig. 1: *Case N. 1.* - Age: 7-day-old male newborn 1989. Teratoma of the head: temporo-zygomatic region developing inside the temporal muscle.

Histology:
a) and b) Components of the 3 germ layers: In a) *Relationship between the temporal muscle and the removed mass.

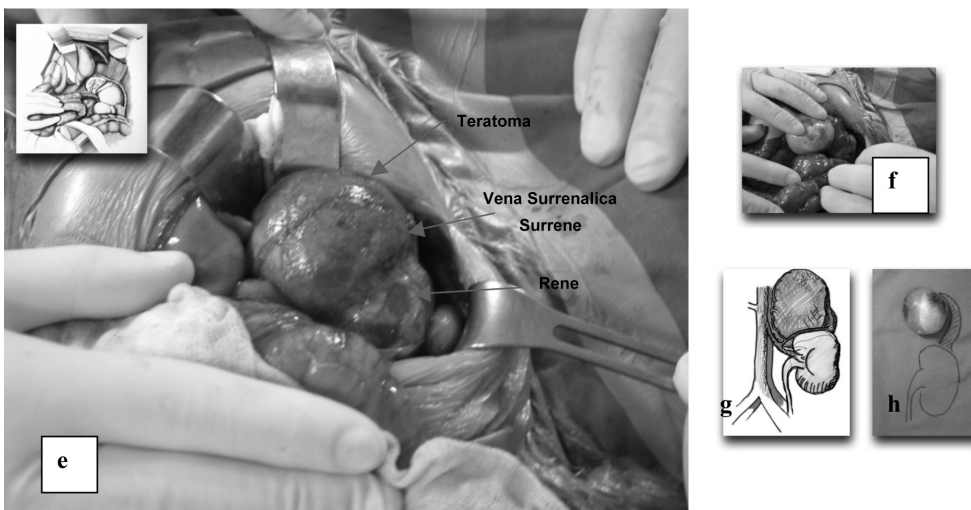
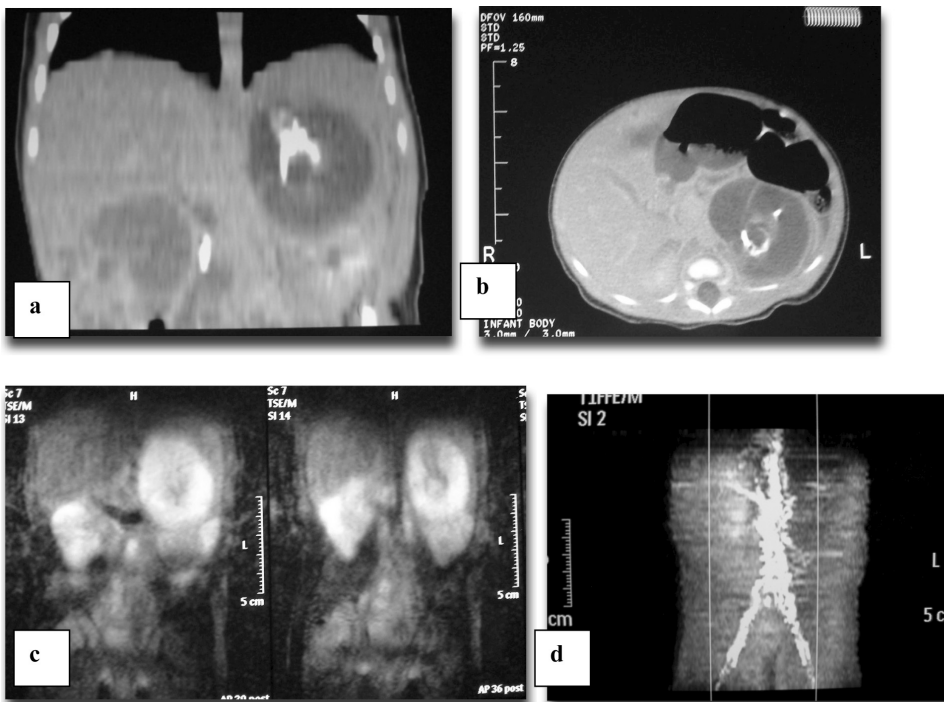


Fig. 2: *Case N. 2.* - Age 28-day-old female newborn 2005. Teratoma of the left retroperitoneal space:

a-d) CT and AngioMRI: Homogeneous predominantly solid non calcified expansile process in the homolateral space, located medially to the adrenal gland displacing downward both the homolateral renal vein and the kidney; e) Operative finding; f) Mass partially isolated from the adrenal gland and kidney; g) Drawing of operative finding; h) Removed mass and sketch of its anatomical relationship with the kidney and adrenal gland.

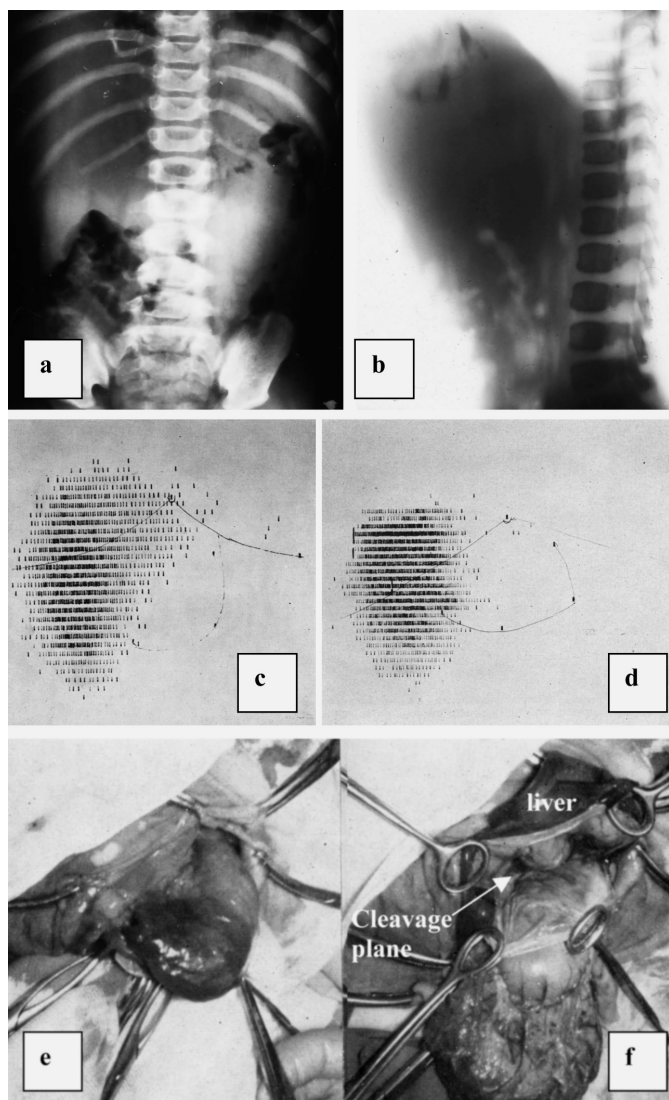


Fig. 3: Case N. 3. - Age 7-month-old female infant 1970, "Fetiform" tripyllic teratoma of the liver

a,b) Abdominal plain Tomography: Organoid calcifications inside the upper pole of a medium density opacity at the abdominal upper quadrant; c, d) Hepatic radio-isotope scan with Au198: large cold area occupying the left lobe of the liver; e) Operative finding: left lobe invaded by the mass, f) Evidence of the cleavage plane after incision of the glissonian sheath.

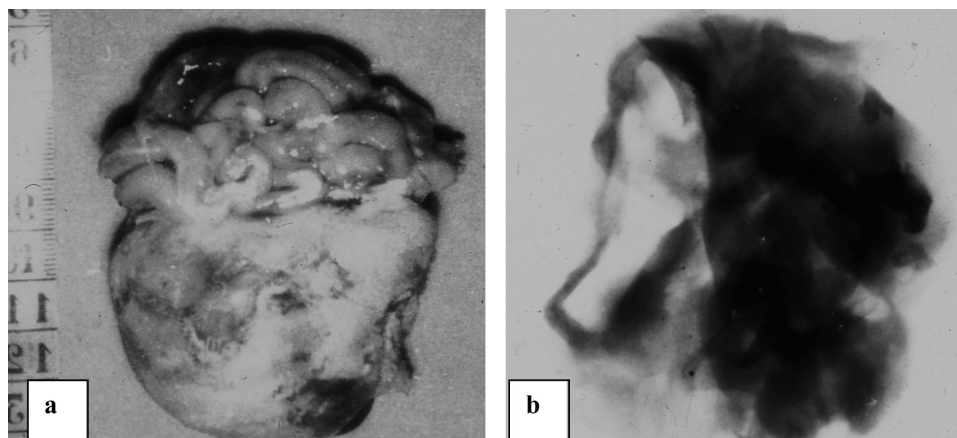


Fig. 4: Case N.3. - Age 7-month-old female infant, 1970

a) Removal Mass: the solid part located in the upper pole of the lesion shows on its surface a twisting encephalic structure delimited by 3-4 mm deep tracks; b) Plain X-ray of the mass: round lobulated heterogeneous opacity containing a twisted tape-like structure, as well as a tooth-like inclusion.

high neutrophilic leukocytosis (WBC 20,000/mm³, N 78%) with elevated values of inflammatory markers (ESR and CRP) only in case N°4. The values were normal in all the other patients. AFP (Table I) was elevated in all the 4 children.

Imaging Studies: The selection of imaging studies was dependent on the clinical presentation and time of hospitalization. In fact, 3 cases (N° 1, 3, 4) were referred and managed many decades ago (1989, 1970, and 1976, respectively). Imaging details are reported in Table II. In any case the imaging studies, consisting mainly of plain radiographies, CT and MRI, detected teratoma and the extension in all the cases, showing evidence of calcification inside a prevalently homogeneous mass, except in case N°2 (2005), where calcifications were missing (Fig. 2 a-d). In infants N°1 (1989) and N°2 the imaging confirmed the prenatal US findings. Hepato-radio-isotope scanning with Au ¹⁹⁸ was performed in addition to plain abdominal radiography (Fig. 3, b; Fig. 3c, d) in case N° 3 (1970) and i.v. pyelography in case n° 4 (1976), before emergency hospitalization (Fig. 6 a, b).

Treatment : With the exception of case N° 3, emergency surgical management was considered advisable in all the other patients, either because of a sudden volumetric increase or because of abscess formation (Case N° 4), as well as the general risk of finding immature tissue or actual malignant transformation from the usually benign type.

The location and extent of the expansile lesion dictated the surgical approach, which consisted of: median laparotomy in case N° 3 (xypho-umbilical laparotomy), transverse supraumbilical laparotomy in cases N° 2 and N° 4. In case N° 1, a low-based skin flap with subcutaneous areolar tissue was created on the right tempo-parietal region to better expose the lesion. In all 4 patients, complete excision was the procedure carried out (Table II). To prevent local recurrence the temporal muscle had to be removed in case N° 1 (Fig. 1a) and the left kidney from which the mass appeared to originate in case N° 4 (Fig. 7). The kidney was rescued in case N° 3 (left Retroperitoneal Teratoma) because the lesion was

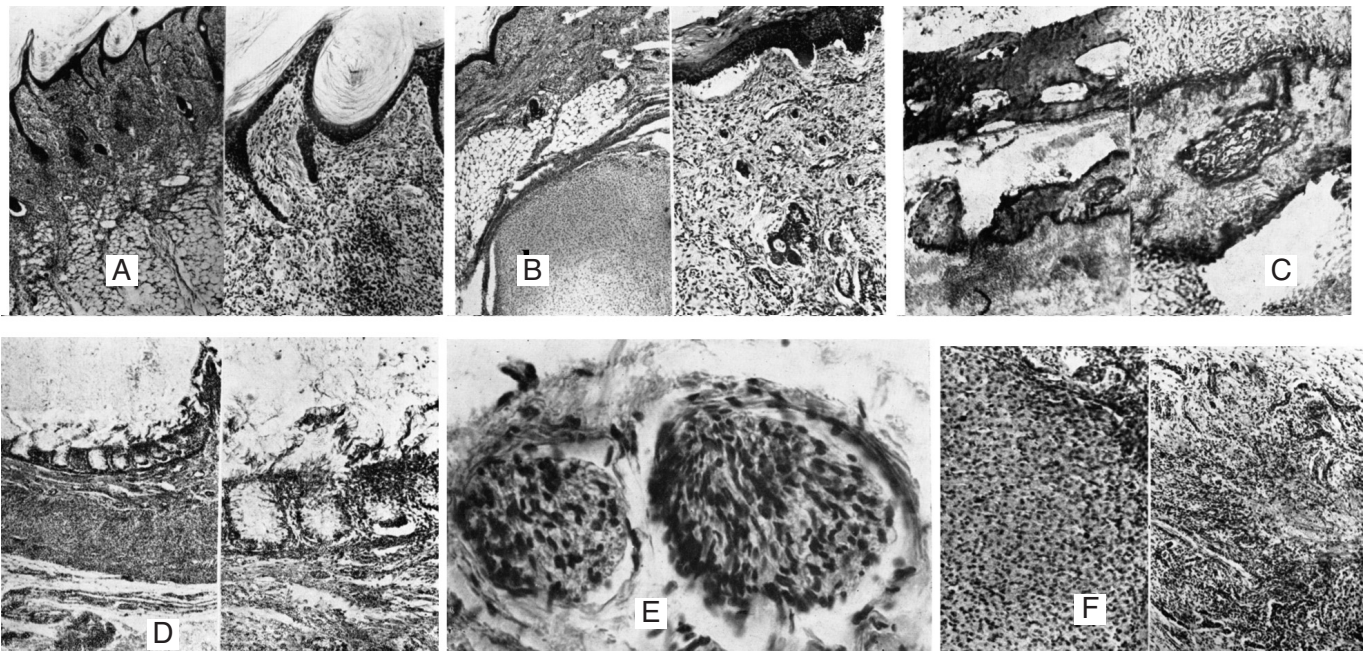


Fig. 5: Case N. 3. - Age: 7-month-old female infant 1970.

Histology: a, b) Stratified squamous keratotic epithelium with glandular appendage, adipose tissue; c) Area of osseous tissue; d) Adult almost normal enteric tubular structures lined by mucous epithelium with intestinal glands; e) Neuro-epithelial areas; f) Close hepatic parenchyma with signs of fibrosis, aspecific inflammation and newly-organized biliary pseudo-tubules.

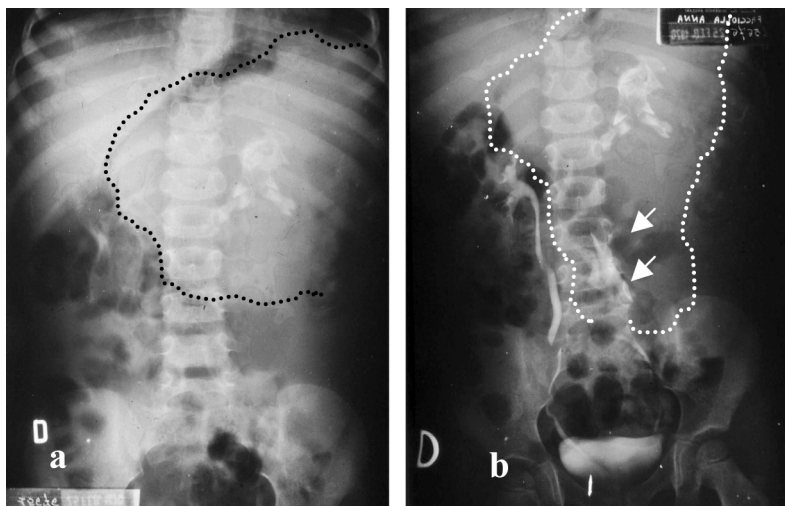


Fig. 6: Case N.4. - Age: 4-year-old female child 1976. "Fetiform" triphyllic teratoma of the left kidney.

Abdominal plain X-ray and i.v. Pyelography: a,b) Calcified jaw-like structure with tooth-like inclusion inside the upper pole of a large solid mass involving the kidney and entirely occupying the left hemi-abdomen overcrossing the midline, the renal excretory cavities (white arrows) being morphologically abnormal and displaced downward and medially. No border between the mass and the left kidney is distinguishable.

easily separable from both the kidney and the adrenal gland (Fig. 2 f-h). In case N° 3 exeresis of the lesion was facilitated by sectioning Glisson's sheath and using a cleavage plane (Fig. 3, f). A small slice of the adjacent hepatic parenchyma was excised for histology. a plain radiograph of the removed mass was taken (Fig. 4 a,b).

Pathology: Based on the gross and histologic features, the site of origin of the tumor was recognized as: the Liver

in case n° 3, left Kidney in case N° 4 (Fig. 7 b, c). Almost all the lesions showed mixed solid-cystic features, but the mass appeared predominantly or almost entirely solid in case N° 4 (Kidney T.- Fig. 7). The final histologic diagnosis was a triphyllic teratoma. The elements of the 3 germ layers were present, but with highly differentiated normal adult-like structures ("fetiform teratomas") in cases N° 3 (Fig. 6 a-f) and N° 4. Some morphological details of cases N° 3 (Fig. 6a-f) and N° 4 are reported in Table III.

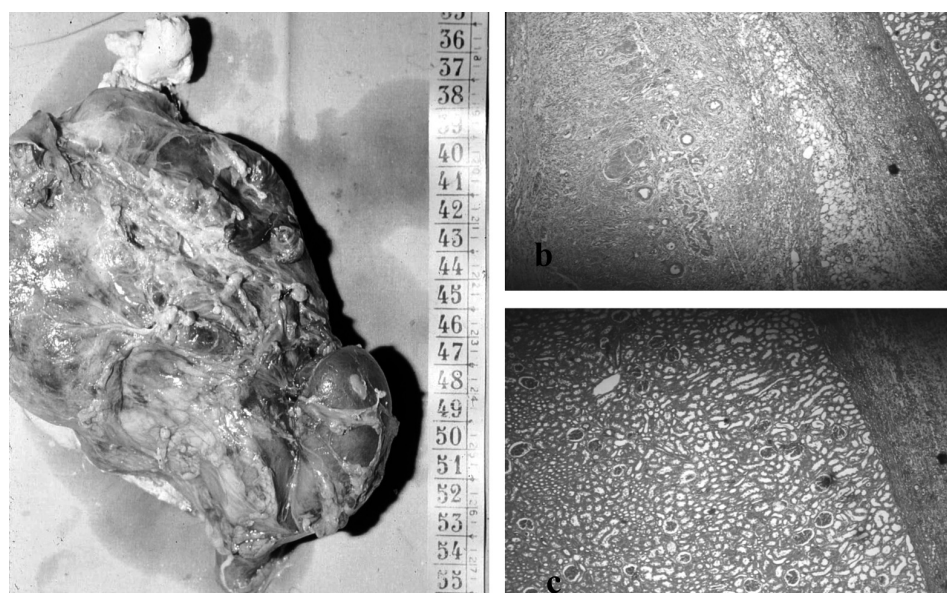


Fig. 7: Case N.4. - Age: 4-year-old female child 1976.

a) Removal mass: Mass: encasing the renal vessels, appears anatomically inseparable from the kidney. b,c) Close histological relationship between the renal parenchyma and the mass, The renal capsule also envelope the neoplastic lesion.

TABLE III - Cases n° 3, 4: Pathologic features

<p>CASE N° 3 Age: 7 mo. F (1970)</p> <p>T. of Liver</p>	<p>GROSS PATHOLOGY</p> <p>Round mixed solid-cystic mass, max Ø cm 9, composed of two portions.</p> <p>Cystic part was partially delimited by a thin, smooth wall, whitish-grey externally and pink inside.</p> <p>Solid part shows on the free surface a twisting encephalic gyri-like structure delimited by 3-4 mm deep linear tracks. Close to the cystic portion small hard osseous swellings are palpable.</p> <p>Mass Radiograph: round lobulated heterogeneous density in which a twisted tape-like structure was contained as well as a tooth-like inclusion.</p>	<p>HISTOLOGY</p> <p>Cystic cavity was lined by typical cutaneous tissue (stratified squamous-keratotic epithelium with glandular appendages, adipose tissue). Inside the wall neuro-epithelial structures were present. The mesenchymal matrix includes areas of cartilaginous and osseous tissue intermingled with adult almost normal enteric tubular structures lined by mucous epithelium with intestinal glands.</p> <p>Hepatic parenchyma: signs of fibrosis, aspecific inflammation with newly formed biliary pseudo-tubules</p> <p>Microscopic Diagnosis: Solid-cystic adult mature "feti-form" Teratoma of the liver.</p>
<p>CASE N °4 Age: 4 y. F (1976)</p> <p>T. of the Left Kidney</p>	<p>GROSS PATHOLOGY</p> <p>Mass including kidney, weight: Kg.1, with different consistency: parenchymatous, osseous, cystic, strident on cutting. Cut surface: organized osseous tissue with tooth-like inclusion and areas of grey solid tissue mixed with cavities containing hair, sebum, dens fetid purulent fluid. A cavity displayed an isolated eye-like structure. It appeared to be impossible to dissociate the kidney from the mass, which occupied the upper half of the organ The renal capsule enveloped both the mass and kidney, in continuity</p>	<p>HISTOLOGY</p> <p>Components of three embryonic layers.</p> <p>Adult high differentiation: skin, bowel muscle-skeletal structures, anlage eye-like structure. Absence of immature or malignant components. The neoplastic tissue appeared beneath the renal capsule.</p> <p>Microscopic Diagnosis: Solid-cystic adult mature "feti-form" Teratoma of the kidney.</p>

Results (Table I)

All 4 patients had an uneventful postoperative course. Clinical surveillance and tests of AFP and other markers were scheduled every 6 months for the first 5 years and annually thereafter. At the current time they are alive, disease-free and have not suffered any recurrence, with a follow-up, as reported in Table I, of: 7 years in case N° 2; 23 years in cases N° 1; 42 years in case N° 3 and 36 Years In Case N°4.

Discussion

Teratomas are embryonal neoplasms of a considerable clinico-scientific and speculative interest. They arise from an abnormal development of the primordial germ cells and / or embryonal totipotent cells. Therefore, they may develop in both gonadal and extragonadal sites. Like gonadal lesions, extragonadal lesions are encapsulated and contain tissues from the three germinal layers. Some display a highly organoid differentiation and these are there-

fore called "fetiform teratomas". They are usually located in the midline, where the totipotent cells arrive by abnormal migration or were primarily included. Rarely, they arise in solid organs (the liver, kidney). Commonly, extracranial teratomas are located in front of the cranial sutures. These neoplasms sometimes exhibit a biologically bizarre behavior. In fact, pure teratomas can have a malignant potential^{1,2}; pure malignant teratomas have shown a tendency to metastasize. The rare form of teratomas with malignant transformation may not contain germinal malignant components but may be of somatic type, such as leukemia, cancer or sarcoma^{3,4}. In addition, a teratoma may present elements of other tumors with germinal cells, particularly YST, even in recurrent primary benign teratomas. These last lesions are considered mixed forms of teratomas; they behave like malignant teratomas and occur more frequently in infants and young children. Finally, a teratoma can be pure and non-malignant but highly aggressive, causing the well-known "Growing Teratoma Syndrome". This syndrome may also appear in the mixed forms after effective adjuvant chemotherapy. It is suspected that chemotherapy is able, in such instances, to destroy the malignant component of the tumor, but spares the pure teratoma, which paradoxically begins to grow very quickly. In any case, extragonadal teratomas are characterized by their rarity and by certain clinical presentations and / or unusual or infrequent histotype which are worthy of note.

CASE N° 1, a 7-day-old male infant, was affected by teratoma of the head, prenatally suspected due to calcification foci, at US scanning. This is an extremely rare lesion which accounts for less than 5% of all teratomas, especially when considering pure extracranial lesions^{5,6}; they are more frequently extracranial extensions of intracranial primary teratomas⁷. Other locations are possible but they generally develop on the cranial sutures, and in 50% of cases, close to the orbit, sometimes eroding the bone. In our patient the site of the lesion was the temporal-zygomatic fossa, on the temporo-sphenoid wing suture, close to the orbit without osseous erosion, and developing within the temporal muscle. Sometimes these tumors appear giant, so large as to occupy and disfigure the cranio-facial region⁸; these are often life-threatening^{8,7}. Microscopically, mature and immature histotypes with YST foci have been reported⁹ (less than 20% - Bernbeck B et al., 2009). Although a case of squamous Carcinoma derived from a cystic mature teratoma of the jaw, AFP-positive¹⁰, is reported in literature, malignant transformation of teratomas of the head is considered rare. Nevertheless, there is a greater tendency to recurrence after incomplete exeresis. Therefore, the gold standard is complete excision, that required the removal of the temporal muscle in our newborn (case N° 4). When the procedure is incomplete, adjuvant chemotherapy should be given, as well as in primary immature lesions⁹. The extensive surgery carried out and the normal AFP values made chemotherapy inad-

visable in our patient, who recovered completely, without recurrence or distant metastases, at 23 years after the diagnosis and surgery (1989).

Retroperitoneal teratomas (case N° 2) account for 1-11% of all retroperitoneal tumors⁽¹¹⁻¹³⁾. The clinical presentation shows 2 peak incidences: infants during the first 6 months and young adults. However, they are infrequent in the pediatric age¹⁴, accounting for just 2-4%¹⁵⁻¹⁷. Under 20 years of age the reported incidence is 70%, distributed as follows: 40% before 10 years, 30% or a little more below 1 year^{14,18-20}. Our case N° 2 was observed and managed when the infant was 28 days old. Excluding cases detected prenatally by ultrasonography, as in our observation²¹⁻²³, the discovery is usually post-natal, and sometimes entirely incidental during diagnostic work-up for an abdominal blunt trauma²⁴ or due to a rapid enlargement of the lesion^{25, 26} or, rarely, urinary retention²⁷ or an abscess complication¹². Generally, they have a benign behavior. However, malignant forms account for 10 -25%^{11,14} with a higher risk in newborns¹⁶ and during the first year of life¹⁷. Terado Y et al. 2010²⁸ describe a rare case of cystic teratoma that occurred in a young adult, containing cells derived from the three germinal layers, but with evidence of an adenocarcinoma arising from well-developed intestinal structures. Some locations¹⁴, or unresectable, otherwise mature lesions, immature and YSTs¹⁶, should be thought to have a malignant potential. Sometimes the diagnosis is very difficult, although the pathognomonic elements are high levels of α -fetoprotein, but not always (Gow KW et al. 2010)²⁹, and calcifications on the imaging study^{30, 31}, even if these are present in only 35-60% of abdominal teratomas³². These elements were missing in our case. Retroperitoneal teratomas are preferentially located on the left side but are occasionally bilateral¹⁶. The location of the tumor may be mistakenly interpreted as the retroperitoneal space during the imaging investigation. Gow KW et al.²⁹ reported a teratoma in a 9-month-old infant, really arising in the retropleural supra-diaphragmatic space but misdiagnosed as a retroperitoneal lesion based on the imaging study alone. A thoraco-abdominal location was also observed in the literature in an infant affected by a congenital diaphragmatic hernia³³; they are usually located close to the kidney and its vessels^{13,17,19}. In the literature, lesions encasing the aorta and the vena cava (Jones et al.2008)¹⁹ have been reported, which pose a challenge during the surgical procedure. Histologically, retroperitoneal teratomas usually present as mature, immature with YST foci or malignant lesions. Rarely, there are nephro-blastic elements inside the lesion. If these are predominant, a differential diagnosis with Wilm's Tumor may be very difficult. There are 6 such cases reported in the literature¹⁵. This particular lesion is still a topic of debate. Cytology is always recognized as a rational tool for the preoperative diagnosis of abdominal tumors, especially retroperitoneal lesions. Nevertheless, cytology has some

limits in the diagnostic work-up of retroperitoneal teratomas. Therefore, the resulting findings must always be compared with the results of imaging, particularly with CT ³⁴. To achieve a better prognosis in managing these teratomas, complete exeresis with preservation of the kidney, as done in our female newborn, is advised as the essential, correct procedure. It is usually technically feasible except in real contingent circumstances ^{17,19}. The literature reports show that immature histotypes, YST, have a tendency to relapse and / or to metastasize, as well as incompletely excised lesions. In such an event, postoperative adjuvant chemotherapy is required ^{35,16, 36}. Our newborn (2005) showed a favorable course and so far she has not had local recurrences or distant metastases, after 7 years from the exeresis. The markers values have also remained within normal limits.

Primary hepatic teratomas, like case N° 3, a 7-month-old female infant, are extremely rare, accounting for less than 1% of cases and, in children, less than 1% of all hepatic neoplasms ^{28,37-45}. They are most commonly observed under 3 years of age ^{46, 47} and are usually located beneath the glissonian sheath or separated from it by a slight rim of parenchyma. Therefore, imaging may be misleading when defining the origin in the liver. However, the hepatic origin of the teratoma may be demonstrated both by hepatic radio-isotope scan with Au 198, as in our infant referred in 1970, and by accurate gross and histologic investigations of the anatomical relationships between the mass/glissonian sheath/parenchyma and extrahepatic ligaments (Fig. 6, f). In addition, pure calcified cystic teratomas may pose some difficulty in differential diagnosis from other acquired and congenital cystic lesions at imaging. In these cases, high levels of α -fetoprotein alone should be a valid aid to diagnosis. Some rare and / or singular pathologic features should be highlighted, such as the association with peritoneal gliomatosis ⁴⁵, the coexistence of a primary teratoma in both the liver and mediastinum ⁴⁸ or mixed lesions in which histopathology may display both mature and YST components in the same neoplasm, clinically associated with a bad prognosis ³⁹, or hepatoblastoma and a malignant teratoma ^{49, 44}, or a primary teratoma with a sarcomatous component ⁵⁰. The rare "feti-form" type in our infant is noteworthy, because of its tendency to differentiate tissues (skin with appendages, neural structures, islets of cartilage and bone trabeculae,) or adult tubular structures such as intestinal tracts provided with typical glands that could be defined as normal. These findings, most frequent in ovarian teratomas of adolescents or adults in the third or fourth decade of life ⁵¹, were also observed in case N° 4. Another important feature is the tendency (4%-8%) of hepatic teratomas to sudden rapid growth, even after the administration of effective adjuvant chemotherapy, if appropriate, and with normalization of the tumoral markers. This event, called the "Growing Teratoma Syndrome", exposes young patients to a high risk of morbidity and mor-

ality ⁵²⁻⁵⁵ and requires emergency, exceptional therapeutic measures such as orthotopic transplantation ⁵⁶. There is, however, an unanimous consensus that complete exeresis provides good results and 100% survival ^{57,58,55}, even in the long term ⁵⁹, while incomplete exeresis favors the development of distant metastases and recurrences ⁵⁵. It has also been demonstrated that adjuvant chemotherapy is effective in both relapsed and immature lesions. The patient we managed with complete exeresis (1970) has remained disease-free and in good health for 42 years. Our experience also includes a teratoma of the kidney in a 4-year-old child (case N° 4). The kidney is statistically shown to be an extremely rare site of teratomas ^{17,60-69}. The first literature report of a neonate was made by GA McCurdy et al. in 1934 (⁷⁰⁻⁸³). In 12 cases reported before 2005 the F: M ratio was 1.8:1. However, a primary teratoma of the kidney may be confused with an extension to the kidney of a retroperitoneal lesion or with a Wilm's tumor showing teratoid features ^{71-75, 67}, especially if the AFP levels are high ⁷⁶. A wide range of congenital anomalies of the kidney and / or of the urinary tract, such as horseshoe kidney ⁷⁷⁻⁸⁰, duplicated collecting system ⁸¹, prune belly syndrome ^{70, 62}, renal dysplasia ⁶⁴, is observed in association with teratomas. Therefore, these malformations may be regarded as a predisposing factor. The most common presenting symptoms and findings are related to the mass effect from a lesion typically located in the retroperitoneal space, or rarely, as in our case, with a complication such as an abscess of a cystic cavity and basal reactive pleuritis. However, the most important diagnostic dilemma is differentiation of a primary teratoma of the kidney from a primary retroperitoneal teratoma with a secondary involvement of the kidney. The renal origin of lesions developing deep within the kidney is easily recognized by imaging investigations. In such instances the following radiological findings can be useful: 1. a peripheral fatty mantle, 2. central attenuation of the density, 3. the "claw sign", 4. distortion of the collecting system. On the contrary, in lesions extensively involving the renal parenchyma with a prevalently exophytic development it may be difficult to define the origin. Radiological findings may be misleading, showing just distortion of the collecting system associated with an apparent pseudo-displacement of the renal shadow due to the mass effect, like in our case N° 4. But the following pathological findings seem very helpful: 1) the kidney is inseparable from the mass both during the surgical procedure and physical examination of the removed gross specimen; 2) the tumoral mass is surrounded by an extension of the same renal capsule, verifiable also at histology. On the other hand, primary teratomas of the retroperitoneal space, apparently infiltrating the kidney, are separable in most cases from the organ during the surgical procedure (^{19, 17}). The abscess formation in the upper pole of the neoplastic mass in our patient did not appear to be the cause of its close anatomic relationship with the kidney.

A purulent inflammation resulted in partial and easily separable adhesions outside the lesion only. Additionally, both gross and histologic examination (Fig. 7 c) confirmed the location beneath the renal capsule with an intraparenchymal development towards the middle portion of the kidney, also involving the main renal vessels at the hilum. Renal teratomas are usually solid and avascular, but sometimes cystic like other cystic renal lesions, from which they must be differentiated^{64,66}. Positive markers are obviously a great help. A rare lesion otherwise detected only in adulthood is Carcinoid Cancer arising inside a mature cystic teratoma, even lacking typical clinical signs of carcinoid syndrome^{83,73,84}. Mature histotypes are prevalent. The grade of differentiation may be so high that some rare lesions are known as “feti-form teratomas”. At histology of the removed masses in our patients, highly differentiated structures were observed, such as normal intestinal-tubular tracts, skin, bone, and even an eye-like structure. But immature⁷⁹ and locally infiltrating or metastasizing⁸² teratomas have also been observed. The histopathologist may have difficulty in excluding, at the diagnostic definition, a renal metastasis of a primary gonadal teratoma or a glomerular and tubular differentiation of a Wilms tumor. A variety of heterologous elements pose the main difficulty^{72,67}. The clinical course is usually benign, but well-differentiated forms can metastasize^{85,68}. The recommended approach is complete exeresis of the mass together with the kidney or partial, usually polar, nephrectomy, but only in anatomically favorable cases, in which a small disease-free rim of renal tissue can be left⁶⁸. Unfortunately, because of their rarity, knowledge about the follow-up of renal teratomas is still limited. Our patient, managed in 1976, has never presented either recurrence or distant metastasis during follow-up lasting 36 years.

Conclusions

From this review of our selected cases and in the light of literature reports, we can draw the following conclusions. Some extragonadal teratomas of childhood may rarely arise in the solid organs (liver, kidney), in the retroperitoneal space or in the cranio-facial region, showing also unique histotypic characteristics (“feti-form”) which distinguish them from more common cases. Being congenital tumors, prenatal diagnosis by US scan is extremely important in order to organize proper perinatal care in appropriate facilities where it is possible to define the diagnosis and carry out emergency surgery. An emergency procedure is frequently dictated both by complications related to the mass effect and the need to define the histology of the whole mass rather than just small biopsy specimens. Some teratomas can hide more or less extensive islands of immaturity or signs of malignant transformation that are clinically evident. It should

be remembered that the high serum levels of α -feto-protein and calcifications on the imaging study, that are usually pathognomonic elements for diagnosis, can be missing in abdominal lesions. Moreover, some additional and specific diagnostic problems can need to be faced by either the radiologist (differential diagnosis from acquired or congenital cystic lesions, identification of the primary site of origin in the liver, kidney or retroperitoneal space) or the histopathologist (exclusion of a renal metastasis of a primary gonadal teratoma or a glomerular and tubular differentiation of a Wilms tumor). The prognosis is generally benign, although AIEOP 2004 guidelines pointed out that high levels of circulating markers, including AFP, in children affected by mature or immature teratomas would indicate the presence of micro-foci of YST, marking them out as at high risk. The UKCCSG II and the SFOP indicated AFP values exceeding 10,000 ng / ml as the threshold identifying a group of patients with a severe prognosis. The treatment indicated is early, complete exeresis, followed by a careful, extensive microscopic examination, associated, if necessary, with adjuvant chemotherapy. Finally, to improve the prognosis, close, long-term clinical, laboratory and imaging surveillance is necessary, at shorter intervals during the first 5 years after the exeresis and annually thereafter.

Riassunto

I teratomi extragonadici sono rari, e, talvolta, presentano aspetti insoliti che li differenziano dai casi comunemente descritti per:

- 1) la sede insolita, (testa, fegato, rene, spazio retroperitoneale);
- 2) la presentazione clinica, a volte acuta, e/o;
- 3) l'istotipo “feti-forme” della lesione. Questi aspetti possono essere isolati o associati nello stesso paziente. Gli Autori hanno estrapolato dall'esperienza complessiva sui teratomi 4 casi insoliti, per lo più operati d'urgenza. Due di essi sono stati trattati subito dopo la nascita.

Scopo di questo lavoro è quello di riportare i dati clinici ed anatomopatologici e di valutarne e discutere l'approccio chirurgico ed il loro comportamento biologico a lungo termine alla luce della sopravvivenza e delle opinioni correnti riportate in letteratura.

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