Hemoperitoneum caused by the rupture of a giant ovarian teratoma in a 9 year-old female.
Case report and literature review

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Hemoperitoneum caused by the rupture of a giant ovarian teratoma ina 9 year-old female. Case report and literature review

The Authors report the case of a 9 year-old girl suffering from acute abdominal pain, combined with mild anaemia (Hb 10.9 g/dL), leukocytosis (24.3 x 10^3 cells/dL), and a large palpable mass in the upper left quadrant. The child underwent an appendectomy 20 days before the admission to our Department. The operation performed in urgency, as well as the removal of a bulky mass situated in the left flank and the right ovary whence it arose, made it clear that abdominal signs and symptoms were caused by the twisting and rupturing of a neoformation, that would hence cause an impressive hemoperitoneum. The histopathologic examination showed a three-germ layer mature mixed teratoma. Clinical, radiologic and biochemical test (α–FP, β–hcG) performed in a postoperative 2 months follow-up revealed no residual disease.

KEY WORDS: Acute abdomen, Hemoperitoneum, Ovarian teratoma

Introduction

Teratomas are nearly the 95% of all ovarian germ-cell tumours [1,2], between 15-20% of all ovarian tumours[3]. We can classify them in three pathologic categories: 1) Mature; 2) Immature; 3) Monodermal [3]. According to Norris Classification Scheme [4], modified by Gonzales and Crussy [5], the maturity of these tumours is established by the amount of immature tissue constituting the mass. Patients affected by immature teratoma have higher rate of recurrence and a worse prognosis than patients with mature teratoma have [6]. Nowadays the most important classification is the WHO's one [6, 7]. Grade 0: mature teratoma with all tissular components well differentiated; Grade I: immature teratoma with less than 10% of immature tissue foci; Grade II: immature teratoma made up of 10 – 50% of immature tissue foci; Grade III: immature teratoma with more than 50% of immature tissue foci [6,7]. Our study reports the uncommon case of a 9-old-child whom undergone an urgent operation because of hemoperitoneum caused by the rupture of a bulky (>20cm) abdominal mass originating from the right ovary. On the histopathologic report this mass has been proven to be a three-layer mature teratoma (Figs. 2 and 3).

Case report

On April 2007 a 9-year-old female child comes to our observation because of a severe abdominal pain persisting for nearly 12 hours with an occlusive symptomatology. The child had undergone an appendectomy in another hospital 20 days before. On admission, no recent or past diseases were reported except for common exanthematic diseases. Pain was widespread on every abdominal quadrant and was continuous, intensified by palpation, with positive Blumberg's sign, and no peristalsis was detectable. On inspection, apart from the scar due to appendectomy, there was a huge convex deformation

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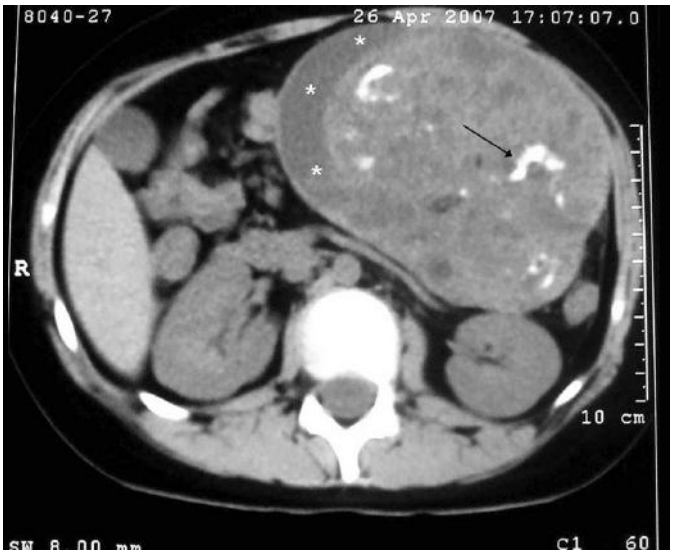

Fig. 1: Image from CT scan showing the bulky mass in the left flank, with its own capsule, with an external liquid portion (white asterisks) and a central solid core including calcified nuclea and little teeth as well (black arrow).

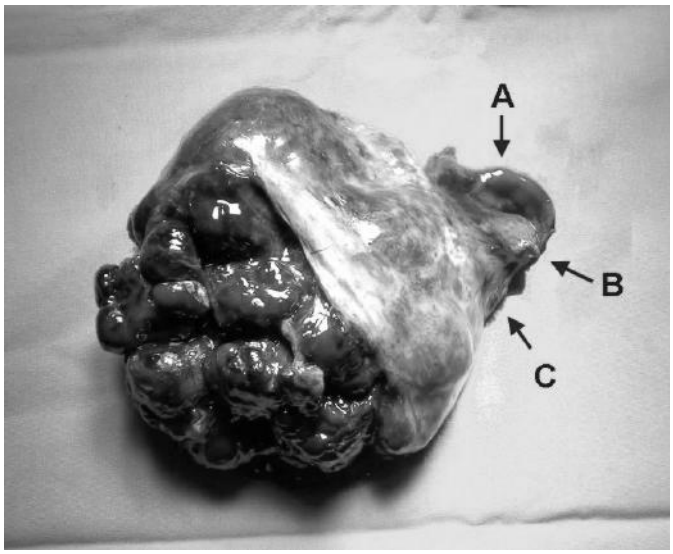

Fig. 2: The bulky mass removed en bloc (14x15 cm), provided with fleshy, hemorrhagic and hard – calcified portions. *A*: ovary and tube; *B*: round ligament; *C*: stalk of teratoma.

of left hypochondrium and flank on the anterior abdominal wall. The mass was hard and elastic, smooth on its surface, not fixed to the anterior and posterior layers. Admission lab tests showed a mild anaemia (10.9 g/dL haemoglobin, $3.84x10^6$ cells/mL red cells, 30,9% hematocrit) and a marked leukocytosis ($24,3x10^3$ cells/mL, 91,3% of which neutrophils). An abdominal CT scan performed without contrast pointed out a large mass in the left flank, mostly provided with its own capsule occupying almost the entire abdominal quadrant, displacing and compressing the closest digestive hallow structures (Fig.1). Moreover, the mass showed a strongly unhomogeneous inner pattern and an external liquid portion surrounding a central core including calcified nuclea and little teeth as well (ARROW). The left renal vein was displaced posteriorly, but it didn't seem to be infiltrated (Fig.1). Hence the patient was soon operated through a median laparotomic sub umbilical - pubic incision. Once the peritoneum was opened, we sucked up 800 cc of evidently hemorrhagic fluid and found out a neoformation of about 20 cm of diameter arising from the right ovary, with its capsule broken in several points, mainly containing blood and provided of a stalk wrung many times on its own axis. Since the stalk rose firmly and widely from the right ovary, we had to perform adnexectomy. Post operative course was normal, except for a heavy anaemia (6,6 g/dL Hb, $2,52x10^6$ cells/mL red cells, 20,3% of Hct) caused by the large amount of blood hold in the mass and in peritoneum. The histopathologic examination proved a three-germ layer mature mixed (solid and cystic) teratoma (Fig 2). Assessment of a-FP and ,b-hCG, and tumoral markers, were dosed only after the operation. We evaluated CEA, a-FP, ,b-hCG, CA125, CA50, CA19-9 on the first day and on the first and the second month after the operation, resulting always in normal ranges. Ultrasonographic examination performed on the 30th and the 60th day after intervention showed no sign of recurrence or remnants of disease.

Discussion

The adnexal torsion is a quite rare event and it often occurs as an acute abdominal pain [8,9]. It is generally caused by a neoplasia or an ovarian cyst, but it can occur in healthy ovaries as well [8,10]. The most common cause of neoplastic adnexal torsion is teratoma (3,5-16,1% of cases) [11,12]. Differential diagnosis may include appendicitis, ectopic pregnancy, inflammatory pelvic disease, endometriosis, diverticular disease, intestinal obstruction, gastroenteritis, pyelonephritis, and renal or ureteral problems [11]. A precocious diagnosis followed by a timely surgical intervention, allows to spare adnexa, especially among fertile women, who have a higher likelihood to develop the disease [11,12]. Ultrasonographic and Doppler examination, and, above all, computerized tomography (CT scan) have been proven essential to detect the ovarian torsion [13,14]. Nevertheless, despite the current diagnostic means, a timely diagnosis is always very difficult, having as a result a delay of the intervention. In a Young Hwan's issue of 1999, a precocious ovary-sparing treatment was possible only in 7% of the cases [11]. Walsh [15] and Kimura [16] highlight the significance of CT for a quick diagnosis of abdominal torsion. They noticed that in these cases the uterus was diverted towards the torsional side, homolateral gonadic vessels were congested, with free abdominal fluid especially in Douglas' space, and that peritumoral fat was stuffed. Young Hwan highlighted that, apart from the signs above mentioned, an

eccentric thickening of the wall of the tumour of 1 cm, along with an enlargement and tortuosity of tubas [11]. Gobel in a report dated back to 1998 writes that surgical therapy alone is the most adequate treatment for the patient affected by a mature teratoma [17]. In a retrospective study based on 58 patients, Norris et al. found a rate of recurrence of 70% for the patients with immature teratoma at stage III [4]. This justifies that the risk of recurrence for these ovarian tumours is strictly related to the histological grading. A similar relationship is not found for immature extragonadal and testicular teratomas. A study conducted in 1999, enlisting paediatric patients with immature ovarian teratomas (82% with stage II-III) described that 97,8% ± 2,8% of them had survived for three years after the surgical therapy alone. The only patient who developed a recurrence survived for 52 months. Extracranial immature teratoma of the paediatric age, with or without malignant elements and over every site and stage, may be treated only with surgery, since the disease-free rate after three years is 93% [18]. In Cushing's opinion, after surgery therapy, the follow-up seems to be enough; in fact patients with recurrence could benefit from a rescue chemotherapy [19]. Moreover, the surgery followed by chemotherapy creates a three years disease-free survival rate between 98% and 100% of cases [18], after being resected again and after chemotherapy [18]. Rescorla suggests chemotherapy whenever -FP is still high after the surgical treatment or after recurrences [20]. Administration of chemotherapeutics might be justified for incomplete resections, where malignant foci could be residual, but it's still not clear if it can avoid the malignant transformations of the mass [6]. Moreover Dark [21] and Gobel [22] say that an appropriate surgical staging for teratoma can be convenient to set the right therapeutic strategy. On the other hand, Lo Curto et al. believe that a watchful strategy after surgical therapy could be suggested for immature teratomas at first stage, but not for those at II and III stage with malignant foci [6]. From what is reported we understand that there's not a singular therapeutic strategy to perform, especially after surgical resection. This is the only mean believed to be necessary and effective [6,18,19]. In our case, since it was a mature teratoma, there was no need to administrate an adjuvant therapy, neither we were doubtful about adopting the laparoscopic or the laparotomic technique. The huge dimension (20 cm), the hard and elastic consistency, and the abdominal rather than pelvic growth of the tumour made us to choose the median laparotomic access, as we believed this way to be the most suitable to remove the mass, as well as for revising and washing abdominal cavity. In fact, since it resulted impossible to empty the neoplasia as suggested by Shalev [23] and came out to be very difficult to put the mass out of the abdominal cavity without a large laparotomy, the laparoscopic technique was considered unsuitable. Several authors think that laparoscopy is not suitable for masses with a diameter larger than 10 cm [23, 24], since Howard, in 1995, suggested the laparotomic approach for tumours larger than 10 cm [25].

Conclusions

The case we report confirms the great effort to diagnose the adnexal torsion by tumour on time. In our experience CT scan wasn't able to point out the ovarian origin of neoplasia, whereas only by the operation and the subsequent pathologic exam we could realize the real kind of neoplasia. We didn't know about any previous ultrasonographic exam of the little patient, so we couldn't know whether neoplasia, definitely already existing at appendectomy, became quickly larger for the congestion produced by torsion, as well we couldn't establish a link between the first operation and the adnexal torsion. We believe that the right therapy for teratoma, and especially for the ovarian one, is based on a multidisciplinary approach; in fact the surgeon, as well as the oncologist, cannot decide a therapeutic protocol without the radiologist or the pathologist, in order to extend the patient's disease-free period.

Riassunto

Riportiamo il caso di una bambina di nove anni che presentava dolore addominale acuto associato a lieve anemia (Hb 10,9 g/dL), leucocitosi (24,3 x 10 cells/dL), e una voluminosa massa palpabile nel quadrante addominale superiore sinistro. La paziente era stata sottoposta ad appendicectomia 20 giorni prima del ricovero nel nostro reparto. L'intervento, eseguito in urgenza, dopo la rimozione di una voluminosa massa situata nel fianco sinistro e originatasi dall'ovaia destra, ha mostrato che tutti i segni e i sintomi addominali erano stati causati da un imponente emoperitoneo dovuto alla torsione e lacerazione della neoformazione stessa.

L'esame istopatologico ha rivelato la presenza di un teratoma maturo trifillico a componente mista solida e cistica

Gli esami clinici e radiologici e biochimici (a-Fetoproteina, b-HCG) eseguiti nei mesi successivi all'intervento non hanno rilevato alcun residuo di malattia.

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