

## CASTLE tumours of the thyroid Value of multiplanar imaging acquisition



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### CASTLE tumour of the thyroid. Value of multiplanar imaging acquisition

**BACKGROUND AND OBJECTIVE:** *Thyroid carcinomas thymus-like differentiation (CASTLE) are rare. The aim of this paper is to address a valid diagnostic and therapeutic protocol for these tumours using three exemplary cases and the previously published literature,*

**METHODS:** *The routine use of Magnetic Resonance Imaging (MRI) in all cases of unclear thyroid fine needle aspiration cytology (FNAC) or in cases of multiple neck masses.*

**RESULTS:** *Three cases of thyroid CASTLE tumours, finally confirmed by histology, are evaluated. All three patients remain in good conditions after an aggressive surgical approach followed by radiotherapy*

**CONCLUSIONS:** *MRI should be routinely used in cases of unclear FNAC or multiple neck masses, Surgery and radiotherapy are effective to manage thyroid CASTLE tumours*

**KEY WORDS:** CASTLE, Magnetic Resonance Imaging, Thyroid tumours.

### Introduction

Thyroid carcinomas showing thymus-like differentiation (CASTLE) are rarely reported<sup>1-5</sup>. Thus, standard diagnostic and therapeutic protocols have not been established. Due to their features, pre- and preoperative evaluation of these tumours is difficult by imaging technology or even fine needle aspiration cytology (FNAC) or frozen sections. A definite diagnosis can only be obtained after surgical excision. We describe the clinical presentation, diagnostic features, surgical approach, and final outcome of three cases in Italy.

### Patients and methods

Since 1988, 477 thyroidectomies have been performed at our Institution and 143 of those were performed due to cancer. A definite preoperative diagnosis could not be obtained in 29 cases. Final histological examinations revealed rare tumours in three of those patients.

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The first patient was a 70-year old smoking male with chronic pulmonary obstructive disease (COPD). He was admitted to our Department in November 2000 with a painless neck mass. The patient had lived with the lesion for two years already. Three months prior to admission the mass had increased in size. The patient complained of dyspnea and dysphagia. A fixed mass involving the right side of the neck was discovered by physical examination. Multiple lymph nodes were palpable. Preoperative otolaryngology (ORL) confirmed paralysis of the right vocal rope. Ultrasound discloses a tumour of 4 cm. in the diameter. It was indistinguishable from adjacent tissues and invaded the neurovascular bundle of the right side of the neck. The internal jugular vein could not be identified, lymph nodes were detected in the Baretz space and near the trachea. The oesophageal wall was thickened and appeared to be of reduced calibre.

Chest X-rays confirmed the oesophageal compression. Thyroid fine needle aspiration cytology (FNAC) revealed a poorly differentiated thyroid cancer. Thyroid functional parameters were within the normal hospital range. The tumour marker levels and preoperative FNAC results are summarized in Table I.

The patient was referred for a neck and chest Magnetic Resonance Imaging (MRI) evaluation. It confirmed the presence of a soft density thyroid tumour involving their right lobe of the gland (Fig. 1). Multiple nodes were

TABLE I – Patient parameters

Patient	CEA nhr 0-7 ng/dl	Thyreoglobulin nhr 0-25 ng/dl	FNAC
Patient n. 1	22	225	Poorly differentiated cancer
Patient n. 2	7	20	Spindle cell lesion
Patient n. 3	6	18	Spindle cell lesion

Legend: CEA = Carcinogenic embryonic antigen  
nhr = normal hospital range  
FNAC = fine needle aspiration cytology.



Fig. 1: MRI showing the involvement of the right thyroid lobe and neck in the first patient.

distributed from the right supraclavicular fossa to the upper mediastinum. They involved the neurovascular bundle and displaced the trachea and esophagus. Tumor tissue was also present within the right and left paratracheal spaces. Additional nodes were detected above the right bronchus. Our therapeutic approach is summarized in Table II.

The second patient was a 62-year old woman. The occasional smoker had been operated due to breast cancer ten year prior to admission to our department. All follow-up examinations had been normal. On admission she complained of dyspnea and mild dysphagia. Physical examination only revealed a palpable enlarged and thickened left lobe of the thyroid gland. ORL evaluation was negative. However, the enlarged left thyroid lobe (7 cm in length) was confirmed by compression and lateral displacement of the trachea and oesophagus. The functional parameters of the thyroid were within the normal hospital range. Table I summarizes the tumour marker

levels and preoperative FNAC results. Preoperative MRI uncovered a partially substernal enlarged left thyroid lobe. The neurovascular bundle appeared to be displaced but not enclosed by the mass. Table II summarizes our therapeutic approach. Surgery revealed a left lobe that appeared completely engrossed by a nodular fleshy mass. Although sternotomy was unnecessary the Lahey maneuver had to be performed to free the thyroid.

The third patient was an asymptomatic 35 year-old man. A mediastinal enlargement was detected during a routine occupational chest X-ray check-up. Neck examination failed to disclose an enlarged thyroid although ultrasound revealed a single nodule (2 cm) within the left lobe of the gland. Preoperative data of the patients are summarized in Table I. MRI confirmed the presence of a single lesion within the gland. Neck lymph nodes were undetectable. However, multiple nodes of small dimension (<2 cm) were detected in the upper mediastinal level. The two findings appeared to be unrelated clinical problems in the beginning. However, the FNAC results and the experience gained with the first patient led us to perform an aggressive surgical approach followed by radiotherapy (Tab. II).

TABLE II – Surgical procedures and outcome

	Surgical approach	Outcome
Patient n. 1	Total thyroidectomy, bilateral supraclavicular and laterocervical lymphectomy, right jugular vein dissection, sternotomy, upper mediastinal lymphectomy Postoperative 50GY radiotherapy	4 days in the ICU, 7 days routine surgical observation, transient hypocalcaemia, right monolateral laryngeal palsy
Patient n. 2	Total thyroidectomy, central compartment lymphectomy	4 days of routine surgical observation, hypocalcemia
Patient n. 3	Total thyroidectomy, central compartment lymphectomy, sternotomy, upper mediastinal lymphectomy Postoperative 50GY radiotherapy	2 days in the ICU, 5 days of routine surgical observation

Legend: ICU = Intensive Care Unit.

## Results

The specimen of the first patient macroscopically resembled a poorly differentiated lesion. Necrotic areas were visible near the fibrous tissue. However, histological examination led to a final diagnosis of a CASTLE tumour (Fig. 2) with cervical and upper mediastinal nodal metastases.

Immunohistochemical methods tested positive for cytokeratins 8, 18, 19, CD5, CEA, and thyreoglobulin but were negative for calcitonin. In Table II we report the surgical outcome of the patient. Twenty months after

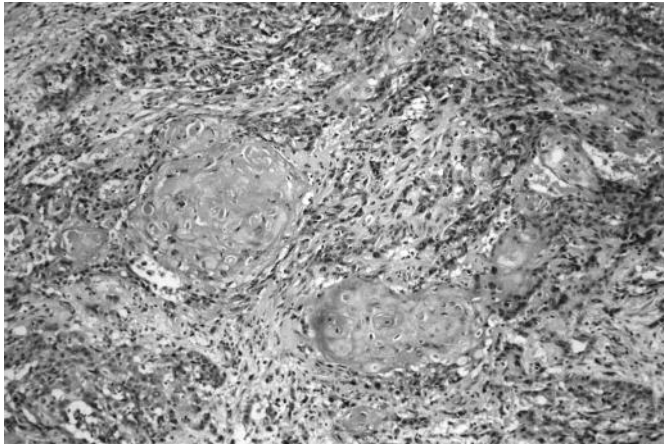


Fig. 2: Histology of the lesion. Evidence of some squamous round areas reminiscent of Hassal's corpuscles.

the intervention the patient was readmitted to our department with dyspnea and dysphagia. MRI showed a mediastinal recurrence that has been managed satisfactorily with radiotherapy.

The specimen of both the second and the third patient stained positive at immunohistochemistry for cytokeratins 8, 18, 19, CD5, and negative for CEA, thyroglobulin and calcitonin. CASTLE tumours of the thyroid were diagnosed histologically in both cases. The two patients are now followed up in 6 month-lasting intervals. Blood tests measuring thyroglobulin and CEA, chest X-rays, and neck ultrasound examinations are routinely performed. We requested an annual check and chest MRI. Both patients seem to be in good condition 22 an 14 months after the intervention, respectively.

## Discussion

Some neoplasms growing within the thyroid gland may mimic the histological features of thymic tissue<sup>1-5</sup>. First descriptions of thyroid tumours showing thymus-like differentiation<sup>6-8</sup> were followed by a classification established by Chan and Rosai<sup>9</sup>. These neoplasms are rare and their biological behaviour remains unclear. A final diagnosis can only be obtained after surgery using immunohistochemical staining techniques<sup>10,11</sup>. Thus, the major problems is correct diagnostic planning in combination with the right therapeutic approach. In other words, it need to be decided whether an aggressive surgical approach has to be planned when a final diagnosis is not available at the time and whether postoperative radiotherapy should be started.

The efficacy of preoperative FNAC or even of frozen sections in such cases is controversial<sup>9,11,12</sup> whereas that the CT scans or MRI has been rated valuable<sup>13,14</sup>. Although the few reports of sporadic cases of CASTLE tumours do not allow to establish a standard diagnosis and treatment protocol, our report may lead to some conclusions. First,

although not routinely suggested for thyroid malignancies<sup>14</sup> we support the use of a multiplanar imaging study techniques such as CT scans or MRI during the preoperative evaluation in the case of positive cytology for cancer. This holds true especially when ultrasound shows multiple neck masses. In contrast to ultrasound or conventional X-ray examinations, CT scans or MRI enable us to screen districts such as the mediastinum. Additionally, these imaging technologies allow for differentiation of schwannomas, neurofibromas, tumours of the carotid body, or secondary tumours of unknown origin<sup>12,13,15</sup>. Moreover, MRI offers the advantage of being a non-invasive, non-toxic technique. It does not require ionising radiation or iodine contrast<sup>16</sup>, which could interfere with eventually necessary radioiodine administration in case of differentiated lesions<sup>14</sup>. Second, if faced with an FNAC diagnosis of poorly or de-differentiated neoplasm involving the thyroid gland we do not support the standard utilization of frozen sections. They can be inconclusive or lead to a wrong diagnosis<sup>12,17</sup> because of the need of an appropriate immunohistochemical staining of final sections. This may lead to an inadequate surgical approach. Finally, patient suffering from CASTLE or other tumours originating from thymic tissue, appear to benefit from wide surgical resection followed the conventional radiotherapy<sup>6,9,12</sup>. We concur with these conclusions.

## Riassunto

**OBIETTIVI:** I carcinomi della tiroide simil-timico (CASTLE) sono rari. Lo scopo di questo lavoro è quello di essere un valido e terapeutico protocollo per questi tumori, con l'analisi di tre casi e della letteratura pubblicata in precedenza.

**METODI:** L'uso di routine della MRI in tutti i casi di non chiara FNAC o nei casi di masse multiple al collo.

**RISULTATI:** Sono stati analizzati tre casi di tumori di CASTLE, confermati dall'esame istologico. Tutti e tre i pazienti sono in buone condizioni dopo intervento chirurgico aggressivo seguito da radioterapia.

**CONCLUSIONI:** MRI deve essere eseguita di routine in caso di non chiara FNAC della tiroide o nel caso di presenza di masse multiple nel collo. La chirurgia ree la radio-terapia sono risultate efficaci nel trattamento dei tumori CASTLE della tiroide.

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