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CASE REPORT

Malignant triton tumour of the anterior mediastinum as incidental finding

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ABSTRACT: ***Malignant triton tumour of the anterior mediastinum as incidental finding. C. Zisis, S. Fragoulis, D. Rontogianni, G. Stratakos, I. Bellenis.***

A rare case of malignant peripheral nerve sheath tumour with rhabdomyoblastic differentiation (malignant triton tumour) of the anterior mediastinum in a 30-year-old male is reported. The tumour was an incidental finding during the diagnostic work-up following a motor vehicle accident. The patient underwent median sternotomy with a tumour resection performed. Local relapse was suspected one month later, as per the chest CT-scan, and post-operative chemoradiation was applied, which produced a response. Twelve months later the patient is doing well while radiological findings remain invariable. Localization of a triton tumour in the anterior mediastinum is extremely rare, adjuvant treatment is necessary, recurrence frequently occurs and the prognosis is dismal.
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Keywords: Mediastinum, mediastinal tumour, neurogenic tumour, surgery.

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Malignant triton tumour (MTT) is a rare malignant neoplasm, of mixed histological character containing both neurogenic and rhabdomyoblastic components. It is considered a histological variant of malignant peripheral nerve sheath tumours (MPNST) with rhabdomyoblastic differentiation. In the majority of the reported cases, the tumour is located across peripheral nerves, close to the spine, in the head and neck region, as well as in the upper and lower extremities.

Case report

A 30-year-old male with a negative medical history presented to the emergency department after a motor accident. He was hemodynamically stable (blood pressure 140/70 mmHg, heart rate 80 bpm), while the clinical examination revealed facial bruises and nasal hemorrhage. The patient complained of chest discomfort and chest x-ray examination revealed a well circumscribed mass in the antero-superior mediastinum retro-sternally. With the suspicion of a retro-sternal hematoma, he underwent a thoracic Computed Tomography (CT) scan, which showed a 6.5 cm mass in contact with the posterior surface of the sternum, not well delineated from the pericardium and the major mediastinal vessels (figure 1).

A median sternotomy was performed, and a multi-lobulated white encapsulated mass was revealed in the mediastinal fat and thymic tissue, with no macroscopically visible pericardial or vascular invasion. A second round, white mass with the same morphology and consistency, 5 cm in maximal diameter, adjacent to the pericardium and the aortic root, was also found, while both phrenic nerves were macroscopically free of invasion. We proceeded to the surgical excision of both masses including the thymus and the whole anterior pericardium. The patient recovered well and was discharged five days after the operation.

Histologically, the tumour was an extreme cellular spindle cell neoplasm with abundant mitosis. Antibody immuno-reactivity to S-100 protein was negative with LEU-7 faintly positive. Some rhabdomyoblastic-like polymorhic neoplastic cells with strong positivity to desmin, myoglobulin, and Myo-D1 antibodies appeared among spindle cells. Such features are compatible with a MPNST with rhabdomyoblastic differentiation i.e. MTT (figures 2 a, b).

Although the entire macroscopically visible affected section was removed, local relapse was suspected as per the CT one month later. Chemoradiation was applied and the lesion remains under control 12 months after the operation.

Discussion

Masson first described Rhabdomyosarcoma within a peripheral nerve tumour in 1932, but the term malignant triton tumour (MTT) was used for the first time by Woodruff and colleagues in 1973, who also proposed diagnosis criteria [1]. The tumour predominantly affects patients in their third and fourth decades of life, while nearly two thirds of cases are associated with von Recklinghausen's disease and concern younger patients [2, 3]. Pres-

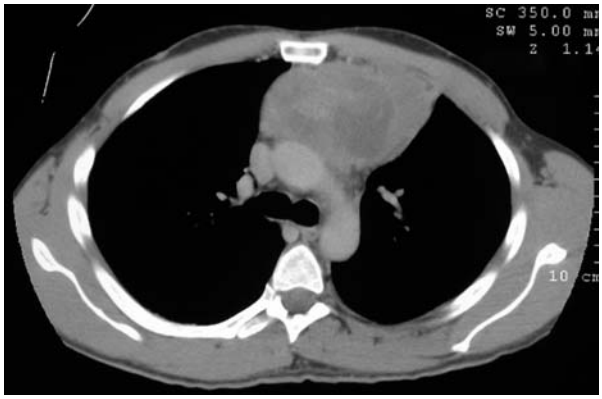

Fig. 1. - Pre-operative chest CT-scan demonstrating the tumor extending to the anterior mediastinum.

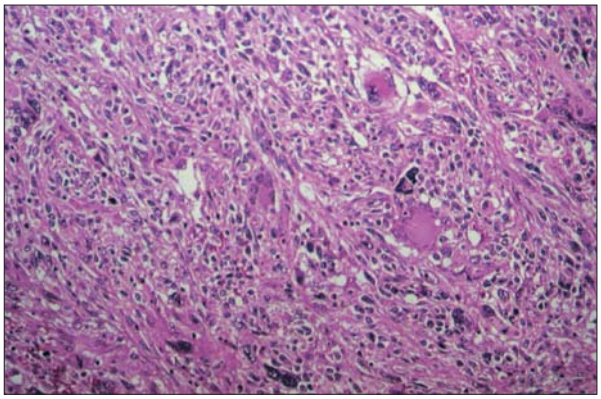

Fig. 2a. - Pleomorphic sarcoma cells. Rhabdomyoblast-like cells in spindle cell area of MPNST. Hematoxylin-Eosin (H+E x 200).

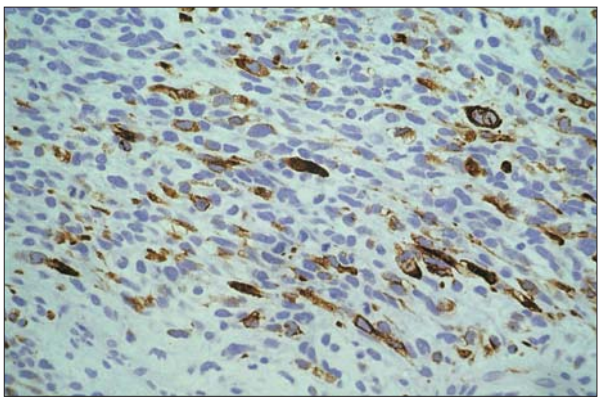

Fig. 2b. - Myoglobulin positive cells (rhabdomyoblastic differentiation) in MPNST. (En vision ABC x 200).

ence in infants and children has been reported, usually relating to genetic alterations and chromosomal aberrations [4,5]; such presence has also been reported in octogenarians [6]. Although such tumours may locate anywhere in the body, one third is located in the head and neck region [7]. Mediastinum location is extremely rare, and Lang-Lazdunski *et al.* have reviewed only 6 MTT mediastinal cases [8]. Among them, two were located in the anterior mediastinum, both of which were submitted to incomplete excision. Another case of MTT, arising from the right vagus nerve and extending from the superior mediastinum to the carinal level in a paravertebral position, has been described with no recurrence signs at 18 months postoperatively [2].

In the case we present, the tumour diagnosis was incidental on the occasion of a motor vehicle accident in an otherwise asymptomatic patient. In the cases sporadically reported in the literature, existing complaints include pain, dyspnea, numbness or sensory deficit.

CT and Magnetic Resonance Imaging (MRI) evaluation do not demonstrate typical imaging features and findings vary depending on tumour location. In the present case, the differential diagnosis included all tumours of the anterior mediastinum, but a slight non-homogenous component within the mass evoked a germ-cell tumour. Percutaneous transthoracic fine-needle aspiration of the tumour under CT does not seem to offer any diagnostic advantage, as the pathology is very rare, the precise nature of the lesion is very difficult to identify without any representative tissue sampling, the risk of inoculation always exists, and the demand for total surgical removal is imperative.

Local recurrence is very common after surgical excision and the reported 5-year survival rate is about 12% [3,8]. Sporadically, favourable prognosis is unexpectedly reported, even after a locoregional relapse, as with an 80-year-old patient, who remains in life 5 years after diagnosis with cerebral involvement and local progression without metastasis [6]. Such a successful outcome has never been reported with a thoracic location of the tumour.

Video-assisted thoracoscopic surgical techniques (VATS) have been used [8], but probably due to misinterpretation of the lesion nature from its preoperative picture. The tumour aggressive behaviour, with its great tendency to recur, establishes the open approach as the preferable access of treatment, in order to achieve a secure resection with wide negative margins. Unfortunately, the tumour demonstrates great invasiveness and its doubling time is highly increased. Furthermore, considering the presence of two masses in the specimen of this patient's tumour, which could not be anticipated from preoperative assessment, we conclude that we have to perform the safest and most extended excision.

Deviation of schwannoma to a triton tumour has been described and malignant degeneration, either spontaneous or induced by irradiation, has been speculated [9,10]. Chromosomal abnormalities reported, regardless of the presence or absence of neurofibromatosis, are probably associated with the poor outcome of the disease [11].

Although the role of chemotherapy or radiation remains undetermined, recommended treatment consists of radical excision, followed by high dose radiation, potentially preventing local recurrence and increasing survival rate [8]. However, successful treatment with intensive high-dose chemotherapy with autologous stem cell transplantation has been reported in MTT of the left masticator space in a 3-year-old boy [4]. Different chemotherapy regimens are tried, but their indication and efficacy remains to be validated.

From this case report we can conclude that such an aggressive tumour as the incidental finding in an otherwise asymptomatic patient is at least uncommon.

The particularity of this entity is characterised by the rarity of its appearance, especially in the mediastinum, as well as its ominous prognosis.

Our knowledge about this pathology has to be improved in order to plan a reliable treatment protocol for patients, and this cannot be too soon. For the moment, the ideal scheme of therapy is practically unknown, due to the small number of cases and lack of adequate experience.

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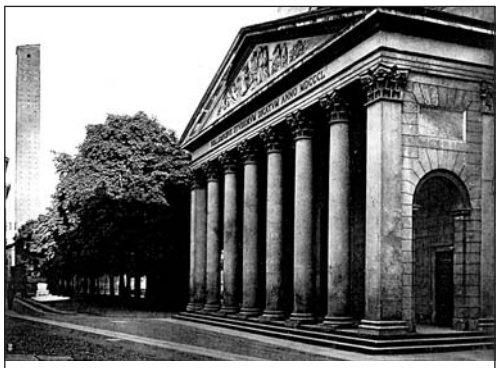

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