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SCIENTIFIC LETTER

Askin tumor: Case report and literature review

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Abstract Askin tumor is an uncommon malignant neoplasm of a neuroectodermic origin that arises from the soft tissues of the thoracopulmonary wall. Defined histologically by Askin and Rosai in 1979 as a *malignant small round cell tumor.* It is described within a group of malignant neoplasms with an aggressive behavior. The lack of clinical guides that establish a standardized management contributes to its poor prognosis and short overall survival. Once a primitive neuroectodermal tumor has been diagnosed, treatment will consist of a multimodal management.

Introduction

Askin tumor is an uncommon malignant neoplasm of a neuroectodermal origin that arises from the soft tissues of the thoraco-pulmonary wall.[1] Histologically designated by Askin and Rosai in 1979 as a *malignant small round cell tumor*,[2] it is described within a group of malignant neoplasms with an aggressive behavior, classified by the World Health Organization (WHO) in 2002 as Ewing's Sarcoma/Peripheral Primitive Neuroectodermal Tumor (SE/PPNETS), organizing tumors of neuroectodermal, bone and soft tissue origin as a single entity. Ewing's sarcoma of the bone, neuroepithelioma, neuroblastoma and peripheral primitive neuroectodermal tumor (PNET), are neoplasms included in this group, and within the PNETs, the Askin tumor.[3]

According to medical literature, there is a greater incidence in younger patients.[1–6] It presents a non-specific clinical behavior, making its precise diagnosis difficult in its early stages.[2–6] Its high rate of local recurrence and the lack of clinical guidelines which establish a standardized management contribute to a less than favorable prognosis and a short survival rate.[4] Due to late diagnoses, large size masses are found, which compromise the tumor's surgical resection because of the presence of adjacent vital anatomic structures.[1] There are, however; studies proving a higher survival rate in patients treated with neoadjuvant chemotherapy followed by a surgical approach and postoperative radiotherapy.[5,6] The contribution made by Demir et al. on surgical treatment clarifies the

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need for a multimodal management, showing that induction with chemotherapy contributes to tumor-free resection margins.[5]

Clinical case

A 25-year-old male with a background of active smoking (12 packs/year), is admitted after presenting intermittent pleuritic pain in the left hemithorax, of variable intensity, of a two-month evolution; accompanied by a non-productive cough, non-quantified fever and a 10 kg weight loss in a course of three months, without a history of occupational exposure hemoptysis or wheezing. Patient refers to have received a previous ambulatory treatment with sulfamethoxazole/trimethoprim, clindamycin and levofloxacin for 7 days without improvement. During physical examination, we observed in a central trachea, cylindrical thorax and symmetric respiratory movements without the use of accessory muscles and with no evident or palpable masses. At auscultation, a reduced vesicular murmur is found in the base of the left hemithorax, with no clinical data added. Blood studies were within normal parameters. Thoracic X-rays are performed, where a radio-opacity is found in the left hemithorax with effacement of the costophrenic angle (Fig. 1). Computed contrasted tomography of the thorax showed the presence of an extra pulmonary tumor (17.6 cm × 12.6 cm × 11.3 cm), dependent on soft tissue of the left thoracic wall, extending from the fourth to the twelfth posterior costal arches, with a heterogeneous rise of the contrast material, without evidence of mediastinal adenopathies (Fig. 2). An incisional biopsy is performed through a left posterolateral thoracotomy with a trans-operatory histopathological report of *a slightly differentiated malignant neoplasia*. Our patient courses with a post-operative evolution with no negative eventualities. After performing a definite histopathological analysis, the presentation of a confirmed primitive neuroectodermal tumor through the presence of small round cells is described (Fig. 3), with immunohistochemical markers positive for FLI-1 and CD99, and negative for cytokeratin, vimentin, TdT, CD45, CD43 and CD56,

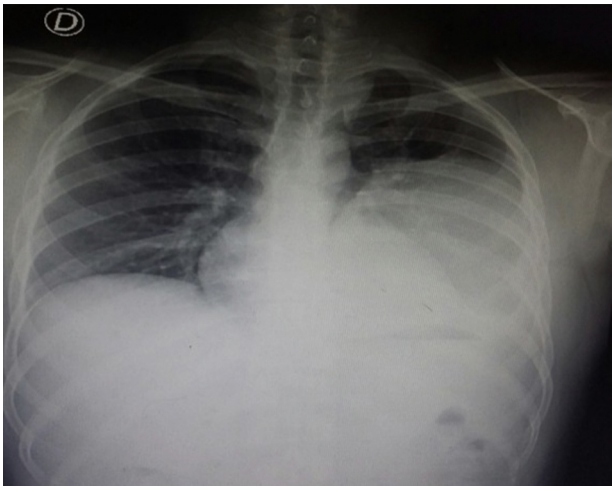

Figure 1 Thoracic X-ray at the time of admission. Radiopacity in the left hemithorax is observed.

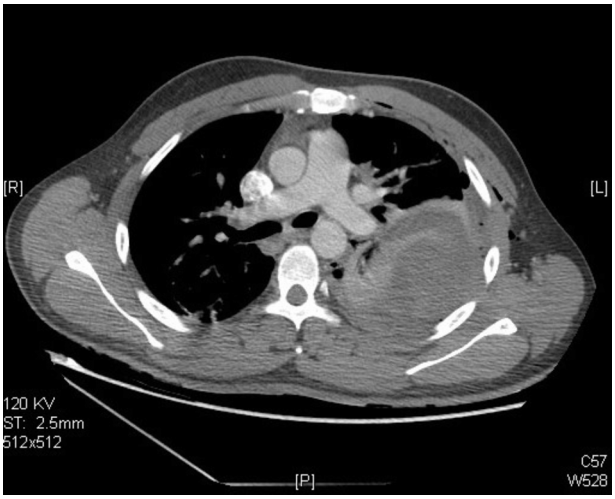

Figure 2 Contrasted thoracic CAT scan. Tumor dependent on the soft tissue of the left thoracic wall.

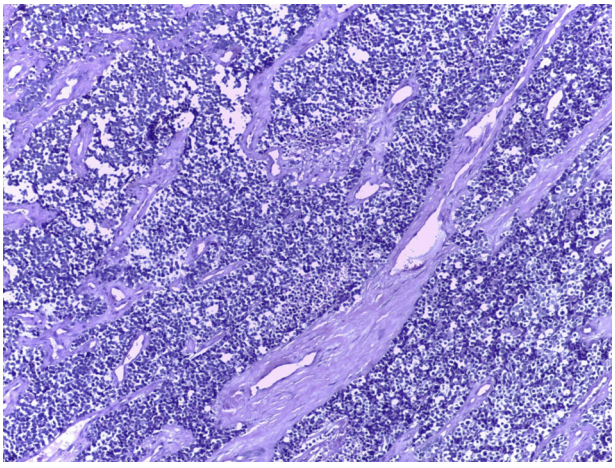

Figure 3 Panoramic view at 5× of the neoplasmic lesion composed of small cells, round and blue, between septa of connective tissue.

with a positive flurosescence *in situ* hybridization (FISH) for rearrangement of the EWSR1 gene (Fig. 4). Oncological management with chemotherapy is started, under an alternate scheme using: vincristine 2 mg/m^2, doxorubicin 75 mg/m^2, cyclophosphamide 1200 mg/m^2 (Day 1) with ifosfamide 1800 mg/m^2 and etoposide 100 mg/m^2 (Day 1–5) every 3 weeks, dosage dependent on patient's tolerance under radiographic control of the tumor. At the time of the edition of this article, the patient is stable, receiving his first cytotoxic cycle, and expecting a reduction in the size of the tumor, thus making resection of the lesion possible.

Discussion

Despite the advent of new diagnostic techniques and tools, Askin tumor has maintained a low incidence since it was first described, which makes us wonder about the factors associated with the onset of the responsible genetic mutation.

Tumors of the SE/PPNET family are categorized under the same entity because of the common histological