

Askin's Tumor in Children: Two Cases Report

Kaoutar Abidi*, Meriem El Baz, Jamila El Houdzi

Pediatric Hematology-Oncology Department, University Hospital Mohammed VI–Marrakesh, Faculty of Medicine and Pharmacy-Marrakesh-Morocco

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*Corresponding author: Kaoutar Abidi

Pediatric Hematology-Oncology Department, University Hospital Mohammed VI–Marrakesh, Faculty of Medicine and Pharmacy-Marrakesh-Morocco

Abstract

Case Report

Askin tumor is a rare malignant thoracopulmonary tumor that primarily affects children. In University Hospital Mohammed VI's Pediatric Oncology-Hematology department, two young patients with Askin's tumor were treated. Chest computed tomography imaging in both patients revealed an Askin's tumor, which was later confirmed histologically. Both patients got postoperative chemotherapy as well as a surgical excision of the tumor. One of them passed away as a result of a localized chest wall recurrence. In this article, we discuss our experiences with this serious pathology.

Keywords: Askin's tumour, Primitive neuroectodermal tumour, Child, Thoracopulmonary region, Ewing's sarcoma.

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INTRODUCTION

Askin's tumor is a primitive neuroectodermal tumor (PNET) of the thoracopulmonary area. It is an uncommon malignant tumor that occasionally develop in the lung and arise from soft tissues of the chest wall. Regarding its individualization and its ties to Ewing's sarcoma, there are still a lot of unanswered concerns [1]. Despite substantial advancements in radiotherapy and chemotherapy, the therapeutic care of Askin's tumor is still not completely standardized. The vital prognosis of these tumors remains very unfavorable because of their metastatic potential and the risk of local recurrence. [2] We discuss two cases of pediatric Askin tumors which were treated at the Pediatric Oncology-Hematology department POH, at university hospital center Mohamed VI in Marrakech.

CASES REPORT

Case 1: A 12-year-old girl, presented a rapidly growing mass in the right omoplate 4 months prior to admission. This mass were painfull with ferm consistancy, measuring 15/8cm. Right parieto-parenchymal mass measuring 14/6 cm was seen on chest imaging (X-ray and chest CT), pushing back the mediastinal structures and associated with pleural thickening. The remaining paraclinical examinations were uneventful. Classified as T1N0M0. The mass was completely removed, and pathology exam revealed a sarcoma-like proliferation of malignant tumor cells that was further supported by an immunohistochemistry finding of positive anti-CD99

and anti-chromogranine A responses. In accordance with the EWING-MEMPHIS protocol, chemotherapy was administered over the course of three stages: an initial phase of treatment lasting five cures (endoxan, adriamycin) and two follow-up phases lasting six cures each (vincristine, actinomycine, endoxan and adriamycin). Treatment ended three years ago, with good improvement.

Case 2: A 13-year-old girl, with no past medical history, who presented with history of left basithoracic pain radiating towards the shoulder with progressive onset for 3 months, associated with exertional dyspnea and a dry cough. The clinical examination revealed a soft left rib mass, mesuring 25/12cm, well limited and painful with an ipsilateral fluid effusion syndrome. The patient had a radiological evaluation ; standard chest X-ray (Figure1), and on the thoracic computed tomography (CT) scan demonstrated (Figure2): heterogeneous equivalent tissue mass mesuring 20/13cm with areas of necrosis continuing at the level of the soft tissues of the left posterolateral wall of the thorax with destruction of the fifth rib and atelectasis of the left lower lobe with an encysted effusion at the level of the left costovertebral gutter suggesting primarily an Askin's tumor. After ruling out other distinct diagnoses by immunohistochemistry, including the presence of anti-CD56, anti-Vimentin, and anti-CD99, anatomical analysis of the parietal biopsied ponction confirmed the histological type in the presence of a tumor proliferation

with small round cells. There were no metastases found in the extension examination and the tumor was classified as T2N0M0. The patient was first, undergone chemotherapy according to mamphis-protocol. CT scan after induction showed progression of volume of tumor

measuring 33/16/18 cm with local infiltration. The second line treatment was done with Euro-Ewing protocol. After the second chemotherapy cycle, the patient experienced localized chest wall recurrence and eventually died.



Figure 1: Chest x-ray showing radiopacity in the left hemithorax

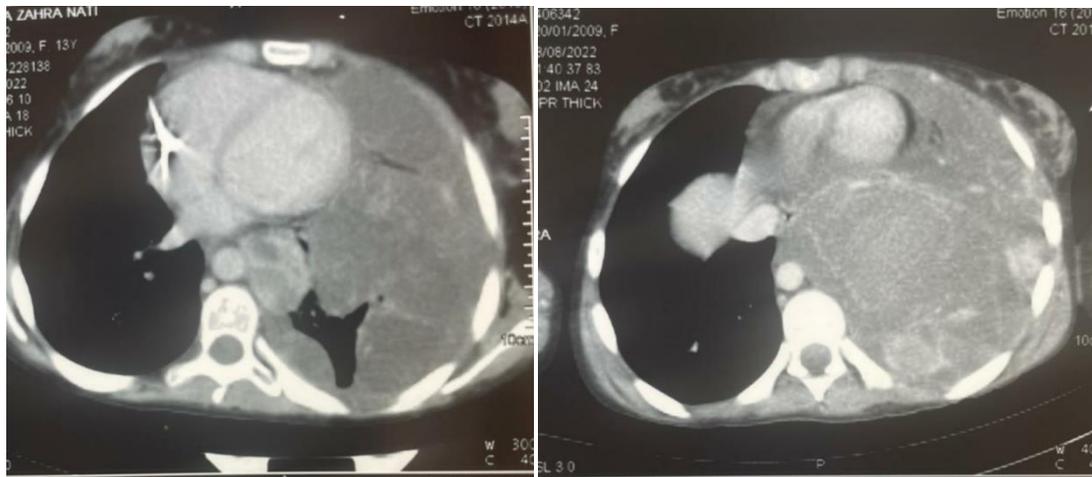


Figure 2: thoracic computed tomography (CT) scan demonstrated a tumor dependent on the soft tissue of the left thoracic wall

DISCUSSION

Primitive neuroectodermal tumor (PNET), which includes peripheral primitive neuroectodermal tumor, neuroblastoma, and primitive neuroectodermal central nervous system tumor, is a type of aggressive tumor that belongs to the Ewing's sarcoma family. Peripheral PNET in the chest wall, also known as Askin's tumor, is one of them and was initially described by Askin *et al.* in 1979. Although it can develop at any age and frequently affects kids and

teenagers, roughly 75% of cases concern females. [1] It first appears as a painful parietal mass that invades soft tissues, the rib cage below, and frequently the lung parenchyma. The most typical radiographic symptom is a chest wall soft-tissue density mass, which can occasionally be accompanied by rib erosion and/or pleural effusion, as in the index case. [3]

The typical CT image of a patient with an Askin tumor shows a heterogenous mass that originates

in the chest wall and has areas of necrosis and bleeding, heterogeneously hypoechoic on ultrasonography, and T1 isointense and T2 hyperintense to muscles with heterogenous gadolinium enhancement on MR. Because a pseudocapsule is present, the masses are often noncalcified and appear well-demarcated. [4] Histologic and immunohistochemical investigation are used to determine the disease's diagnosis. The tumor's cytologic smears show that the malignant cells are tiny, spherical, poorly cytoplasmic, and grouped in rows. Homer-Wright rosettes with different layers of cells and fibrillary material are a common finding. The tumor exhibits positive immunohistochemistry results for a number of neural markers, including NSE, CD99, and vimentin. [5]

Other solid tumors, like peripheral primitive neuroectodermal tumors, should be taken into consideration in the differential diagnosis in any child presenting with musculoskeletal symptoms, despite the fact that other illnesses, like leukemia, lymphoma, and neuroblastoma, may be accompanied by musculoskeletal manifestations in children. [6] Combination therapies, which include chemotherapy, surgical tumor excision, and radiation therapy, offer better prognosis outcomes for patients. Relapses, on the other side, frequently take place and necessitate more intensive treatments, including high-dose chemotherapy, monoclonal antibodies, and bone marrow transplantation. [7]

Historically, RT plus adjuvant treatment with VACA (vincristine, dactinomycin, cyclophosphamide, and doxorubicin) performed better than VAC in Intergroup Ewing Sarcoma Study trials (IESS-I and IEES-II) (vincristine, dactinomycin, and cyclophosphamide). Trials conducted afterwards showed that leaving out dactinomycin had no discernible effect on clinical results. Several studies assessed the effects of including ifosfamide and etoposide in conventional chemotherapy. Patients with standard-risk were included in the Euro-EWING99-R1 trial to determine whether cyclophosphamide may substitute ifosfamide in consolidation therapy. The results showed that VAC (vincristine, dactinomycin, and cyclophosphamide) was statistically similar to VAI (vincristine, dactinomycin, and ifosfamide). The methods of local control treatment include surgical resection and RT. The effectiveness of these two

strategies has not yet been tested in any trials. However, it was discovered that surgery combined with RT was linked to a decreased risk of local failure [7,8].

CONCLUSION

Askin's tumor is a particularly rare and aggressive type of tumor. The prognosis for children with this disease is still dismal, and the treatment protocol has not yet been standardized. For patients with or without metastases, the standard of care entails multimodal chemotherapy and local therapy, which includes surgery and radiotherapy (RT).

CONFLICT OF INTEREST

None.

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