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# Pancoast and Tobias Syndrome Revealing Costal Ewing's Sarcoma about a Case

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**Abstract** **Case Report**

Pancoast and Tobias syndrome is usually caused by lung cancer. However, other causes can be described as in the case of our observation, hence the interest of histological confirmation in order to institute an adapted treatment.

**Keywords:** Pancoast and Tobias syndrome, Ewing's sarcoma.



## INTRODUCTION

Ewing's sarcoma (ES) is a primary malignant bone tumor of children and adolescents and more rarely of adults after 30 years of age [1]. It can affect all bones of the skeleton, with a slight preference for the long bones (50-60% of cases) and the lower extremity. Flat bones are frequent locations, mainly the pelvis and more rarely the ribs [2]. Costal SE is a very aggressive tumor characterized by a high potential for local recurrence and distant metastasis. The introduction of a multimodal approach including chemotherapy, surgery and radiotherapy in the management has led to improved local tumor control and survival [1, 2]. We present an observation of costal SE with the diagnostic and therapeutic approach of this tumor.

## OBSERVATION

We report the observation of Mrs. RB, 38 years old, with no particular pathological history, admitted for cervico-brachial neuralgia associated with a left supra-clavicular swelling, progressing for 6 months in a context of altered general condition and apyrexia. The physical examination revealed a Claude-Bernard-Horner syndrome (enophthalmia, narrowing of the oculo-palpebral cleft and miosis), a collateral venous circulation of the left hemithorax, an exquisite pain on palpation of the left anterior thoracic wall in front of the 1st and 2nd ribs and a hard and fixed left supra-clavicular mass of 5 cm of large diameter. The chest radiograph (figure 1) showed a dense, homogeneous opacity of the left apex with costal lysis of the anterior and middle arches of the left 1st rib.

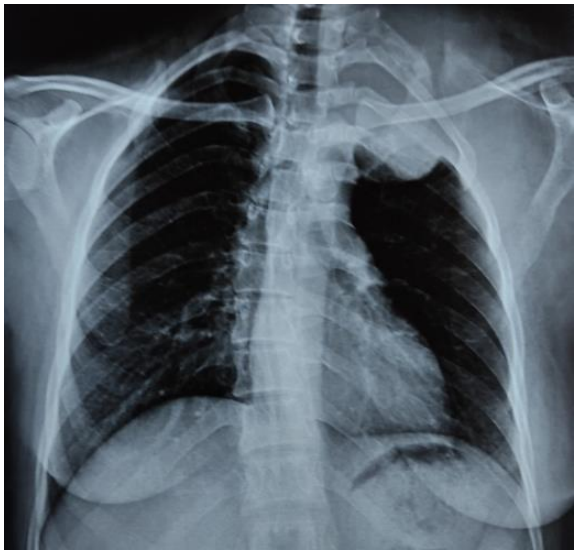


**Figure 1: Front view chest radiograph showing a dense, homogeneous left apex opacity with lysis of the anterior and middle arches of the 1st rib**

The thoracic CT scan (Figure 2) showed a left parietal tissue mass with exo and endothoracic development, centered on the first two left ribs with lysis of these ribs, coming into contact with the vertebral bodies from C7 to T3 without invading it and heterogeneously enhancing after injection of contrast medium.

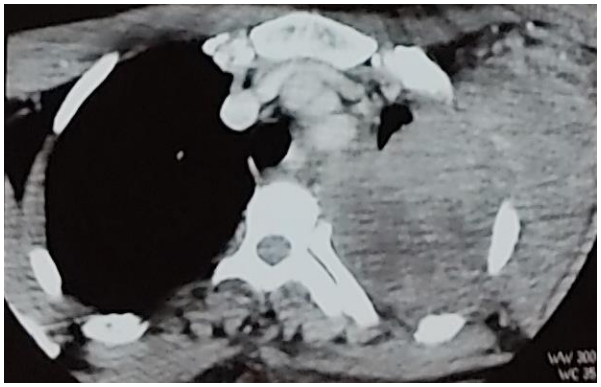


**Figure 2: Axial section thoracic CT scan showing a voluminous left parietal lesion process with exo- and endothoracic development centered on the rib with rib lysis**

Thoracic magnetic resonance imaging (Figure 3) did not reveal any endocanal extension. The diagnosis was confirmed by transparietal ultrasound-guided biopsy. The microscopic aspect showed a malignant tumor proliferation arranged in layers, with round cells. The immunohistochemical study showed diffuse expression of CD99 and the genetic study showed a rearrangement of the EWSR1 gene.

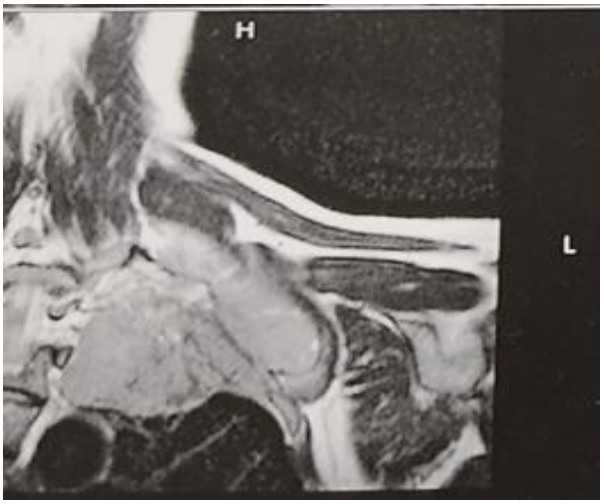
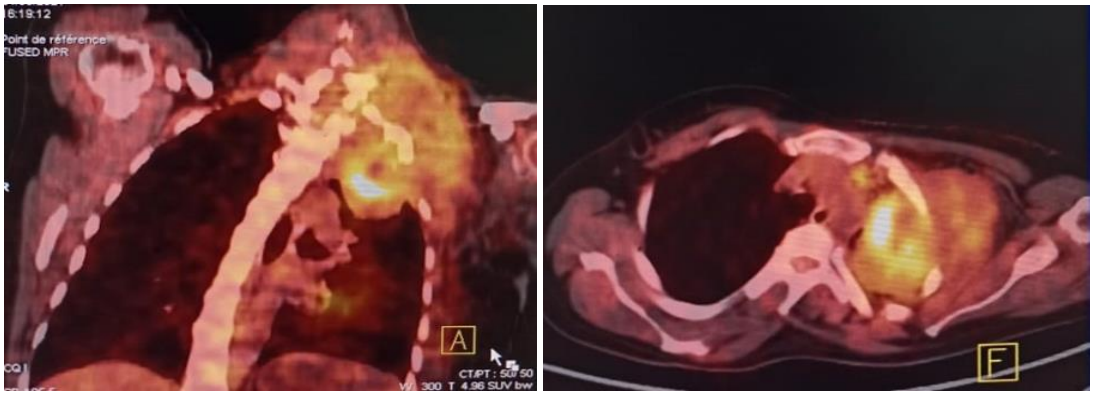



**Figure 3: Cervical MRI showed the lesion process pushing back the left pulmonary apex and without spinal extension**

The PET scan (Figure 4) showed a hypermetabolic left cervicothoracic mass invading the first two left ribs, hypermetabolic homolateral mediastinal adenopathies with no secondary distant location.




**A** **B**

**Figure 4: PET scan in axial and sagittal section illustrated a hypermetabolic left cervicothoracic mass invading the first two left ribs with aortopulmonary window adenopathies. A: Frontal section. B: Axial section**

The patient had received 6 courses of chemotherapy based on Vincristine, Actinomycin, Cyclophosphamide, Ifosfamide and Etoposide. A clinical and radiological re-evaluation after 6 courses of chemotherapy showed a regression of the volume of the mass.

## DISCUSSION

Ewing's sarcoma is a rare tumor, first described by James Ewing in 1921 as a tumor of neuroectodermal differentiation. It represents 5-10% of malignant costal tumors [2, 3]. It preferentially affects male subjects of Caucasian origin [4].

Currently, Ewing sarcoma is thought to originate from mesenchymal stem cells, and the EWS-FLI-1 fusion protein is thought to be responsible for the differentiation of mesenchymal stem cells into Ewing sarcoma stem cells that have phenotypic characteristics of neural crest cells [5].

In most cases, it presents as a parietal mass, chest pain, often associated with fever and asthenia. Respiratory symptoms (cough, dyspnea) or pleural effusion are also possible [6]. Sometimes, unusual signs may be associated such as isolated Claude Bernard-Horner syndrome or Pancoast-Tobias syndrome as in our observation [6].

Ewing's sarcoma is often accompanied by a large mass, indicative of aggressiveness, round or oval, heterogeneous in appearance, with ill-defined contours, and eccentric to the rib, with exo- and especially intrathoracic development, which may exert a

significant mass effect on the mediastinum [7, 8]. MRI is still indicated for vertebral and/or posterior costal involvement in order to evaluate the endo-canal spinal extension [3].

Histological confirmation is performed either by percutaneous biopsy or by surgical biopsy, which is the reference method for the diagnosis of primary bone tumors and the most efficient sampling method with a risk of non- contributory biopsy of only 7% [9]. The macroscopic appearance shows a multilobed, soft, friable, and often necrotic- hemorrhagic tumor [10]. Microscopic examination reveals a proliferation of tumor cells arranged in large, rounded trabeculae measuring 12 to 14 micrometers. Their nuclei are oval or rounded and have dense but scattered chromatin (Figure 5). The cytoplasm is pale and very sparse. The immunohistochemical study has two main functions. It allows to rule out certain differential diagnoses and to bring positive elements in favor of the diagnosis of Ewing's tumor represented by a diffuse and membrane expression of the p30/32 protein (CD99) encoded by the MIC 2 gene (Figure 6) [11]. However, CD99 is not specific to Ewing's tumor. It is expressed in many normal cells of the body such as fibroblasts, endothelial cells, osteoblasts, or lymphoid cells, but also in other tumors such as lymphoblastic lymphomas, carcinomas, which does not allow to establish the diagnosis with certainty, hence the interest of molecular biology that confirms the diagnosis, by demonstrating the chromosomal translocation t (11, 12) (q24; q12) (Figure 7), by analysis of chromosomal bands, by fluorescence in situ hybridization (FISH) or by RT-PCR [12, 13].

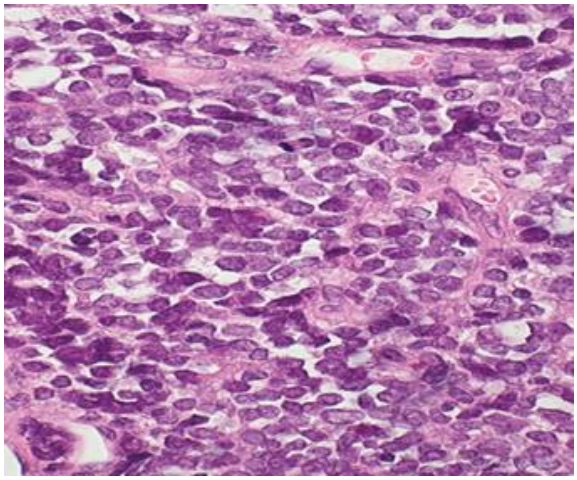


**Figure 5: Microscopic view of tumor cells at high magnification, rounded, with oval nuclei well delimited by a thin membrane. Cytoplasm is extremely reduced in size. Periodic acid Schiff staining (PAS), magnification × 640**

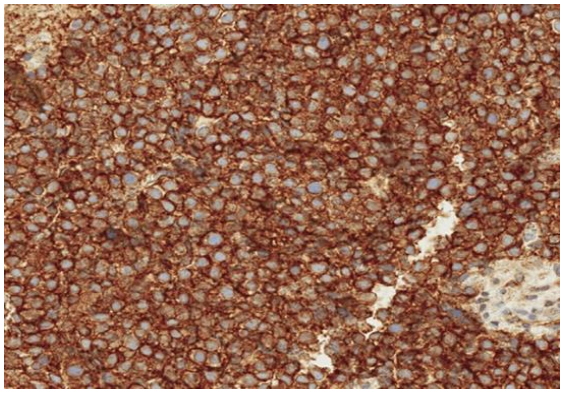


**Figure 6: Typical membrane expression of CD99. Immunohistochemical staining for HBA71, magnification× 400**

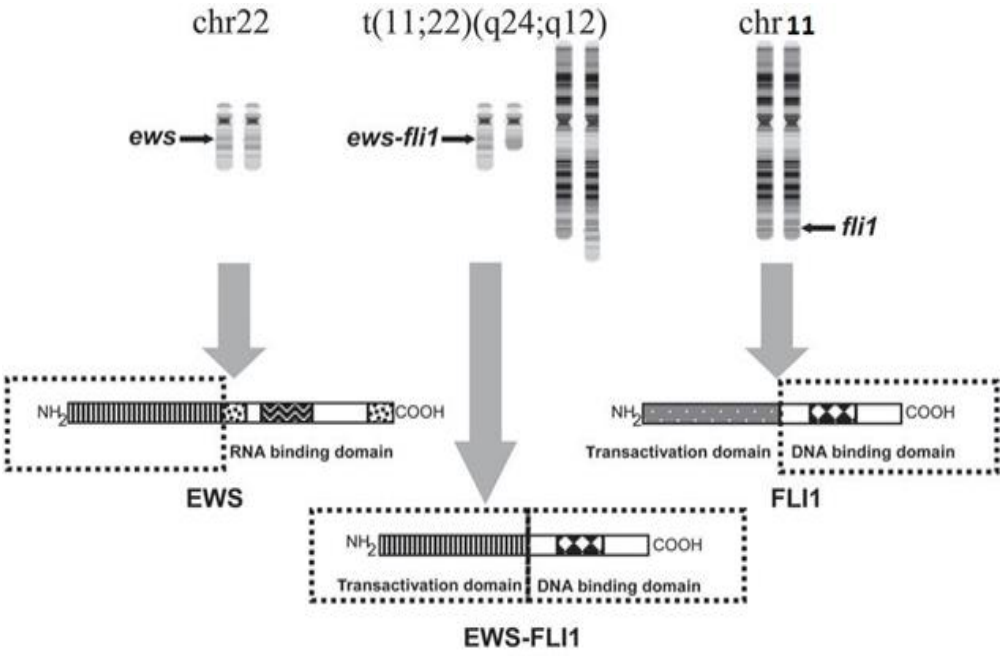



**Figure 7: Schematic showing the reciprocal translocation t (11;22) (q24;q12)**

PET scan, bone marrow biopsy, and brain CT or MRI in case of neurological signs are essential to look for distant metastases, mainly in the lungs, bones and bone marrow and more rarely in the brain and liver [13, 14]. Our patient had no distant metastasis at the time of diagnosis.

Surgical excision is the treatment of choice for localized tumors [15]. Neoadjuvant chemotherapy remains essential to reduce the volume of the primary tumor and treat micro-metastases. Current protocols are based on multidrug therapy (4 to 6 cycles) combining two to six anticancer drugs (vincristine, doxorubicin, cyclophosphamide, ifosfamide, etoposide, with or without actinomycin D) [2]. Radiation therapy is indicated for rapidly progressive and inoperable tumors or when the histological response to chemotherapy has been inadequate. Sometimes it can be combined with surgery to reduce the risk of local recurrence [13, 16]. In our patient, surgery was considered after 6 courses of neoadjuvant chemotherapy.

The evolution is usually rapid, with a tendency to soft tissue invasion and the appearance of distant metastases [13, 14].

Local recurrence after surgical resection is frequent and the prognosis is very poor, depending mainly on the existence of metastases and the response to chemotherapy [13].

The five-year recurrence-free survival is currently 70% in localized forms and 25-30% in metastatic forms [17].

## CONCLUSION

Ewing's sarcoma remains relatively rare, but its localization in the ribs is not exceptional [1]. Its diagnosis is currently based on histology and molecular biology data. Its treatment combines chemotherapy and complete surgical removal of the tumor [2].

### CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

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