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Peripheral Primitive Neuro-Ectodermal Tumor of the Pleura: About a Rare Caser, with Literature Review

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Abstract:- Primary peripheral neuro-ectodermal tumor (pPNET) is a rare and very aggressive tumor that belongs to a small round cell tumor, and is most often located in the chest wall.

PNET of the chest wall belongs to the Ewing sarcoma family because of their genotypic and phenotypic appearance, It can affect several organs, Pleural localization remains rare and little described in the literature. Here, we report a case of a particular localization of a pPNET. This is a 30-year-old patient who presented with chest pain on the right side, posterior irradiation, stage III dyspnea of MMRC and a dry cough, all progressing in a context of apyrexia and significant weight loss. .The chest X-ray showed complete opacification of the right hemi-thorax with deviation from the midline to the left side. Thoracic computed tomography reveals a large right pleural mass of cystic density containing multiple raised septa after injection of the PDC, pushing back the mediastinum to the left, the liver below, and the vascular structures of the neck above and inside which, however, remain permeable, associated a pleural effusion of moderate abundance, without other secondary localizations. The diagnosis of pleural Pnet was made on an echo-guided biopsy of the necrotic pleural mass. The patient was then transferred to the cancer center to start chemotherapy. We insist through this work on the rarity of the pathology, the diagnostic difficulties and the reserved prognosis.

Keywords:-
- *Peripheral primitive neuro-ectodermal tumor (PNET)*
- *Pleural space*
- *Necrotic pleural mass*

I. INTRODUCTION

Primary neuro-ectodermal tumors (PNETs) are rare malignant tumors and currently group together a set of lesions having in common: A morphological appearance of a small round basophilic cell tumor with or without rosettes (resulting from cells of the embryonic neural crest), which mainly occur in children and young adults. PNET occurs outside the central nervous system, and can affect the ribs, sternum, scapula, collarbone, and soft tissues of the chest wall. Pleural involvement is rare. Called primary peripheral neuro-ectodermal tumor (pPNET). Primary peripheral neuro-ectodermal tumors of the chest wall belong to the Ewing sarcoma family due to their genotypic and phenotypic appearance. The PNET of the pleura is a very rare localization, little described in the literature, with a tendency for rapid loco-regional evolution and a poor prognosis. Presenting this clinical case to us to highlight the rarity and aggressiveness of this tumor, as well as the peculiarities of this exceptional pleural location.

II. OBSERVATION

41-year-old young woman, never treated for tuberculosis, without any notion of recent tuberculosis contagion , without any particular history, consults for right chest pain with posterior irradiation which has been evolving for 4 months, associated with exertional dyspnea with progressive worsening becoming stage III of MMRC and an intermittent dry cough, all evolving in a context of deterioration of the general condition and of significant weight loss not quantified Clinical The clinical examination revealed syndrom of fluid effusion from the entire right hemi-thorax. A chest x-ray [Figure1] revealed a homogeneous water-toned opacity occupying the entire right hemithorax with a deviation from the midline to the left side. Thoracic computed tomography [figure2,3] showed a large cystic multilocular right pleural mass containing reinforced septa and thick adenoids after injection of the contrast product with complete right lung collapse pushing mediastinal structures medially and associated pleural effusion. The pleural fluid contained 40 g/dL of protein, and was characterized by a lymphocyte predominance in a hemorrhagic background. An ultrasound-guided biopsy of the pleural mass was performed. Pathological examination revealed the presence of a tumor proliferation consisting of small round cells endowed with a lean basophilic cytoplasm and small rounded nuclei with fine chromatin, finely nucleolus. These cells are placed in a diffuse manner crossed by fine capillaries. The immunohistochemistry conclude has a histological appearance and immunohistochemically profile in favor of a PNET tumor. With diffuse and intense positivity of anti CD99 drugs, while the markers (anti CD20, CD5, CD56, anti chromogramin , anti synaptophysin , anti AE1 / AE3 , anti EMA, anti desmin, anti myogenin) were

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negative. The patient was transferred to the cancer center for chemotherapy.

III. DISCUSSION

In 1979, Askin et al. describes a rare, malignant small cell tumor (Askin tumor) that occurs in the soft tissues of the chest wall, sometimes in the bones, or rarely on the periphery of the lung. This neoplasm is now recognized as a type of primary neuroectodermal tumor (PNET). Askin tumors probably develop from embryonic migratory neural crest cells [1].

Peripheral PNET occurs outside of the central and sympathetic nervous systems and differs from central PNET in that peripheral PNET generally expresses high amounts of the MIC2 antigen (CD99) and exhibits a very characteristic chromosomal translocation.

Peripheral PNET is rare and the overall incidence is 1% of all sarcomas. This tumor can occur at any age, although the maximum incidence by age is adolescence and young adulthood, Overall, 27% of cases occur in first decade, 64% in the second decade and 9% in the third decade of life, while cases occurring later in life are infrequent [2]. The incidence in men is slightly higher than in women (ratio of 1.1) [3]. In general, PNET is a very aggressive neoplasm and its prognosis is poor, with a 5-year disease-free survival rate of 45-55%. [4]

Peripheral PNETs are extremely malignant tumors, and bone and soft tissue are their primary sites. Most commonly, these tumors affect the chest wall, paravertebral region, pelvis, and limbs and are described in a range of organs such as the kidneys, bladder, and heart [5]. Chest wall PNET can affect the ribs, sternum, scapula, collarbone, and soft tissues of the chest wall; pleural localization is less common [6] .Traditionally, distinctions were made between classical Ewing's sarcoma and PNET, it is now accepted that the two are a spectrum of a single neoplastic entity. Peripheral neuro-ectodermal tumor lesions are typically painful, in keeping with the capacity of thoracic tumours to invade the chest wall, lung or mediastinum. They are generally Soft and fleshy with areas of haemorrhage and necrosis.

The initial clinical symptomatology, which sometimes precedes the diagnosis by several months, is most often chest pain (39% of cases) related to a thoracic wall mass.

Sometimes, the diagnosis is made at the stage of complications such as Claude-Bernard-Horner syndrome related to a mass extended to the apex or when lymph node or systemic metastases are discovered.

On the biological level, the search for urinary cathecholamines is constantly negative.

The radiological aspects are not specific to Askin's tumor. Indeed, the parietal mass can be associated with a pleural reaction, in the form of thickening in about 70% of cases or effusion in 28% of cases. The latter is most often scarce, but it is sometimes the only anomaly. Costal invasion is found in about 40% of cases [7].On the other hand, the purely intercostal site of the parietal mass is rare. Hilar or mediastinal lymphadenopathy may be observed. Isolated parenchymal masses are exceptional [8] .The radiologic appearance of Askin's tumor may be that of a metastasis or another round cell tumor. The CT presentation is often in the form of a lump heterogeneous chest wall. Magnetic resonance imaging gives the same images as computed tomography and helps in assessing the degree of mediastino-pleural invasion and exothoracic extension and especially damage to the brachial plexus in the event of associated pain of the upper limb.

The macroscopic appearance of these tumors is that of a gray-whitish mass, often round or ovoid with multiple nodules or multilobulation: this mass generally measures between 2 and 14 cm in diameter with areas of necrosis and hemorrhage when cut. The tumor will present small round malignant cells containing little cytoplasm and are arranged in rows in the cytological smears. Using immunohistochemistry, the tumor is positive for multiple neuronal markers, such as neuron specific enolase (NSE), CD99, and vimentin.

That must be distinguished from other malignant small cell tumors of the mediastinum and chest wall, namely [9]: Ewing's sarcoma, alveolar rhabdomyosarcoma, neuroblastoma, neuroepithelioma and non-Hodgkin's malignant lymphomas .The patient described here presented in the second decade of life with a pleural mass and massive hemorrhagic effusion, and unresectable disease that showed the typical histological and histochemical features of PNET. Given the scarcity and recent individualization of disease, no treatment regimen has been validated in the literature. Patients are most often treated on a case-by-case basis. It seems that the combination of excisional surgery, loco-regional radiotherapy and multidrug therapy be advocated by the majority of authors .Thus, the surgery must be as carcinological as possible because the quality of the excision correlates closely with the risk of local recurrence. Complementary treatment based on radiotherapy or chemotherapy is not currently well codified. All decisions around the therapeutic strategy do not can only be taken as part of a multidisciplinary consultation in a specialized center.

It is important to exclude non-Hodgkin lymphoma from the differential because its treatment does not include surgery. The prognosis for Askin's tumor is very poor. Recurrences localities are very common. In addition, metastases are already present in 10% of cases at the time of diagnosis. The metastatic sites are in order of frequency, the lung contralateral, lymphadenopathy, bone, and, more rarely, the liver, adrenals, brain, retroperitoneum and sympathetic system. Cases of medullary, retro-orbital, retrocrural and oropharyngeal have been reported [10].The prognosis was poor with 2 and 6 year survival rates of 38% and14%. The average survival rate after recurrence is reduced to 11 months.

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Looking at the English literature, few cases have been reported in the plural PNET.

IV. CONCLUSION

In conclusion, the peripheral primary neuro-ectodermal tumor of the pleura is a very aggressive tumor that can occur at any age. And should be included in the differential diagnosis of tumors of the chest wall, especially in children and adolescents, although isolated cases have been described in patients of all ages. Diagnosis requires immune-histochemical workup supported by imaging. Treatment includes chemotherapy, surgical resection, and radiationtherapy. The prognosis of pPNET is poor with a high risk of recurrence

Declaration of interests

The authors declare no competing interest

LISTS OF FIGURES

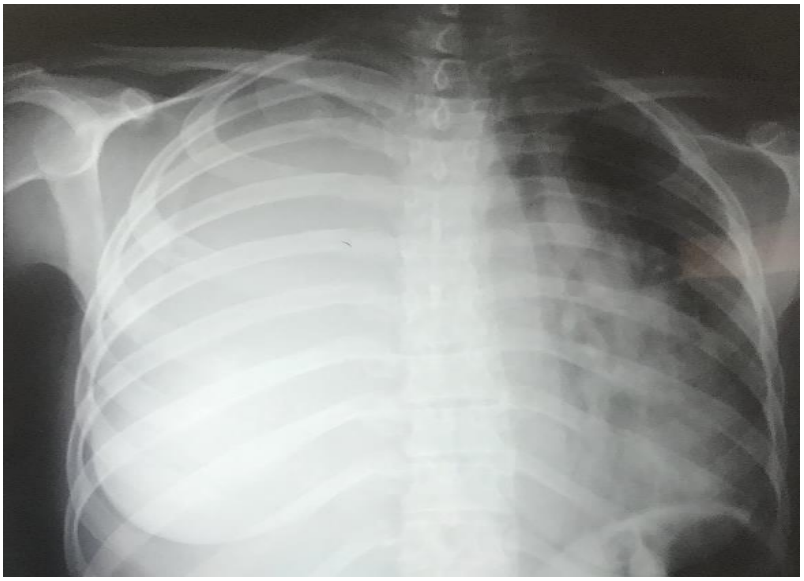

Figure 1: The postero-anterior chest roentgenogram showed a homogeneous opacity occupying the totality of right thoracic field.

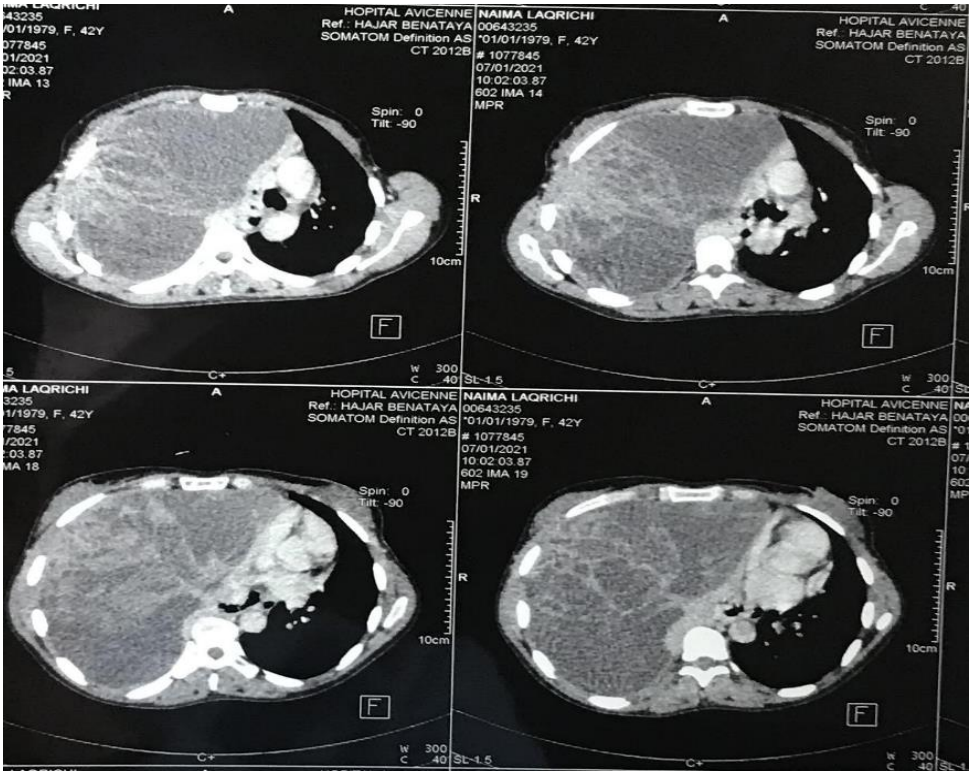

Figure 2 : axial CT thoracic CT shows a large right pleural mass enhanced after injection of the PDC (black arrow) with pleural effusion (white arrow)

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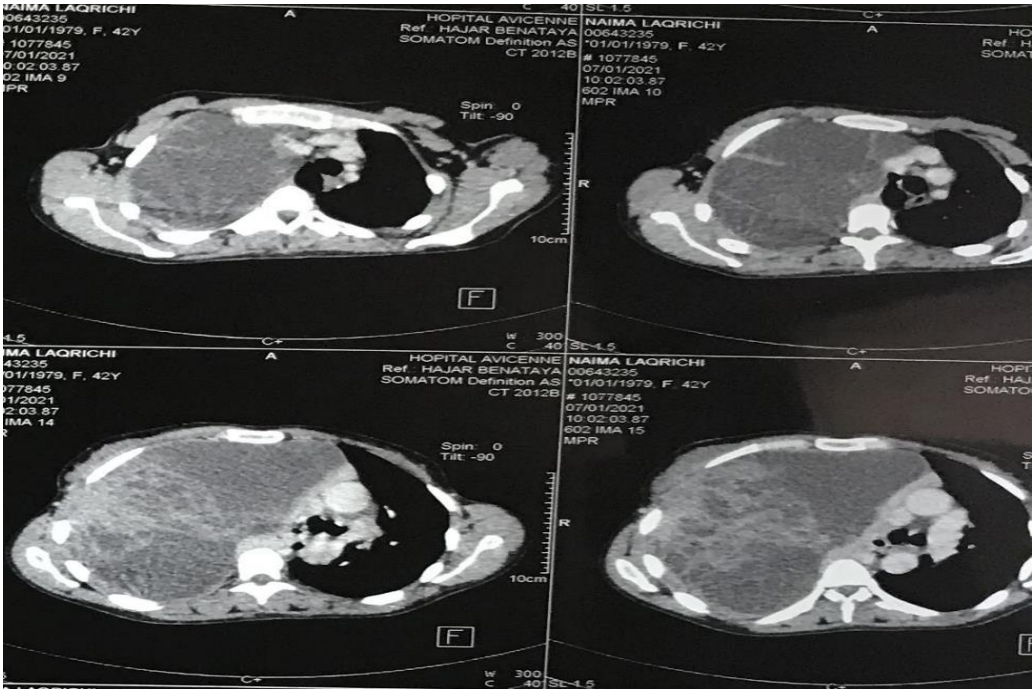

Figure 3 : axial CT thoracic CT shows a large right pleural mass enhanced after injection of the PDC with pleural effusion (white arrow)

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