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Alveolar Rhabdomyosarcoma Revealed By Breast and Lymph Node Metastasis: A Case Report

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Abstract: *Rhabdomyosarcoma (RMS) is a rare malignant tumor of the skeletal muscle. The most frequent localizations are in the head and neck (40%), genitourinary tract (25%) and limbs (20%). There are two types of RMS: the embryonal type, which is more frequent in children, and the alveolar type, which is more frequent in adults and more aggressive. Involvement of the breast, either as a primary tumor or as a metastasis, is extremely rare. We report here a case of primary alveolar RMS of the limbs with breast metastasis in a 17-year-old girl followed in oncology for RMS who presented a breast mass in which the biopsy came back in favor of a breast metastasis of RMS, we will try to give an update on the management of this rare localization of RMS*

Keywords: Rare disease · Alveolar rhabdomyosarcoma · Breast metastasis · poor prognosis

Introduction:

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children and adolescents [1, 2]. Although metastasis is common and can occur in 20% of RMS, particularly in the alveolar histological type, breast involvement is a very rare location, but may pose a problem in therapeutic management [3–5]. Breast metastases of RMS occur almost exclusively in adolescent girls, with a poor prognosis as for other metastatic RMS [6, 7]. To date, there is no consensus on the optimal treatment strategy for breast metastases in RMS due to the limited number of reported case series. We report our experience with a 17-year-old girl with primary alveolar RMS of the extremities with iliac, inguinal and cervical lymph node localization with distant breast localization and present a review of the literature on diagnostic and therapeutic implications.

Case Presentation:
17-year-old girl, without any particular history, her history of HDM disease goes back to 4 months before her admission with the installation of a mechanical pain in the right lower limb after a trauma, 2 months later, the symptomatology worsened with the increase of the volume of the lower limb without inflammatory signs, with the installation of pelvic pain and a skin nodule at the scapular level and cervical ADPs. A cervico thoracic scan was performed showing the presence of several ADPs in the right iliac and inguinal chains, some of which had a necrotic center and the largest of which measured 24 mm in minor axis. Biopsy of the left cervical ADP showed a histological aspect of a malignant round cell tumor, and the immunohistochemical complement was performed, showing anti Bcl2 Ac: focal positivity, anti CD10 Ac: focal positivity, anti synaptophysin Ac: focal positivity, anti myogenin Ac: diffuse positivity, anti desmin Ac: focal positivity, KI 67 at 95%, which concluded in a metastasis of an alveolar rhabdomyosarcoma, the assessment of extension made by a thoraco-abdomino-pelvic scanner objectified a left breast tissue nodule of 40mm in diameter (figure 1), with irregular contours with the presence of poly ADPs of the right primitive iliac chain and of the right internal and external iliac chains measuring 40x30mm for the largest one, a complement by Echo-mammography made having classified the breast nodule BI-RADS 4, the Microbiopsy was made showing an aspect of alveolar rhabdomyosarcoma (figure 5,6). Whole body metabolic exploration by 18F-FDG PET/CT is a powerful tool for the assessment of the extension of these tumors, due to lack of means and its unavailability at the treating hospital, our patient didn't benefit from this examination. The patient was put on 3 cycles of VAC (vincristine, dactinomycin and cyclophosphamide). Concomitant intensity-modulated radiation therapy was initiated 08 weeks after the start of chemotherapy and targeted the primary site and involved regional lymph nodes. with poor clinical and radiological response because the evolution was marked by the occurrence of signs of spinal cord compression with, on spinal MRI, left apical pleural thickening with intracanal extension determining a spinal cord compression at the D2 to D4 levels with signs of suffering of the spinal cord and nerve roots (figure 2,3,4). The prognosis was considered very poor and the family preferred the option of palliative treatment. The patient died of severe respiratory distress 1 month later.

Discussion:
Breast involvement in alveolar Rhabdomyosarcoma is a very rare entity, either as a primary tumor or secondary localization. Seventy cases of girls younger than 19 years of age with breast metastases of RMS have been reported between 1980 and 2014 [3]. Most of these cases of breast metastasis occurred in pubertal patients. The explanation for this phenomenon in post puberty especially remains unclear, but many potential contributing factors have been suggested [6]. The tendency of RMS to metastasize to the breast in adolescent girls is thought to be related to the expression of insulin-like growth factor (IGF) receptors in the neoplastic cells. Breast epithelium and stroma are known to express IGF-I and IGF-II growth factors [8].The rapid increase in vascularity and growth of the mammary gland during this phase of pubertal development is another hypothesis for preferential metastasis in the developing breast

[9]. Yaren et al. even reported a case of metastatic RMS in the breast in a pregnant woman. This reinforced the hypothesis that the vasculature plays an important role in the hematogenous spread of the disease [10].Breast investigations are not routinely requested in the routine diagnostic workup of girls with alveolar RMS. We believe that a careful physical examination of the mammary gland should be performed at the time of diagnosis and during follow-up, and that any suspicious breast lesions should be confirmed by breast ultrasound and MRI [11]. Treatment of patients with alveolar RMS and those with incomplete resection usually consists of chemotherapy followed by concurrent chemoradiation [12]. Although alveolar RMS is a chemosensitive neoplasm, local progression and relapse are often responsible for treatment failure [13]. Radiation therapy has been shown to be effective on metastases, but the optimal dose and fractionation have yet to be established [14].In our patient, treatment was based on CAV chemotherapy combined with radiotherapy [8, 13]. Treatment of breast metastases of RMS is extremely challenging.It is not yet known whether the criteria commonly used in adults can be adopted in young pediatric population. In particular, it is unclear whether surgery should be complete or conservative, and whether irradiation of the breast regions in young prepubertal adolescents is feasible despite the high risk of radiation-induced tumors and growth sequelae.D'Angelo et al recommended a surgical approach in the case of a single breast metastasis and greater use of radiation therapy for patients with multiple breast metastases [4].

Because the breast location of RMS is unusual, its treatment is not well established [7]. For these patients, the inability to eradicate occult microscopic residual disease remains a great challenge. Several cellular pathways appear to be involved in the pathogenesis and survival of RMS. Huh et al. highlighted the possibility of a mesenchymal stem cell as a progenitor of alveolar RMS and the role of high-dose chemotherapy with stem cell rescue in high-risk patients [2]. Whether targeting genetically quiescent cells with longer-term maintenance therapy in patients with minimal residual disease, or focusing on the development of targeted therapies, new treatment strategies are desperately needed if we are to end the well-intentioned but unsuccessful efforts of the past four decades [15].

Conclusion :

We conclude that, although breast involvement in rhabdomyosarcoma is rare, it is important for physicians to recognize it as a potential cause of a painless breast mass in young girls and to keep in mind that this malignant disease is often misdiagnosed as a benign fibroadenoma. Delay in diagnosis and treatment may result in a worse prognosis for the patient, as this tumor is aggressive in nature and has a long-term survival rate of only about 50%. Surgical treatment in this age group can have a devastating impact on the patient's self-image. Therefore, we recommend prompt and complete surgical mastectomy after diagnosis, with adequate breast reconstruction.

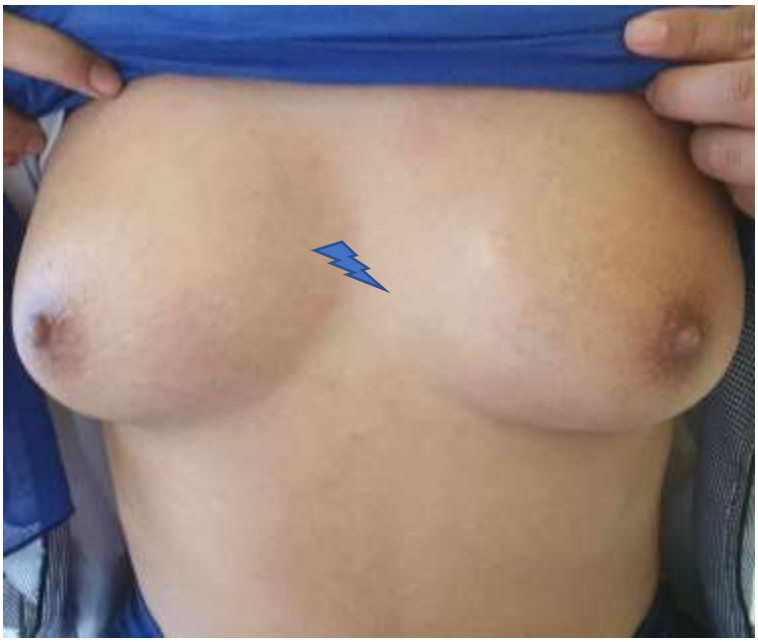

Figure 1: photographic image showing a small lump in the left breast related to alveolar rhabdomyosarcoma

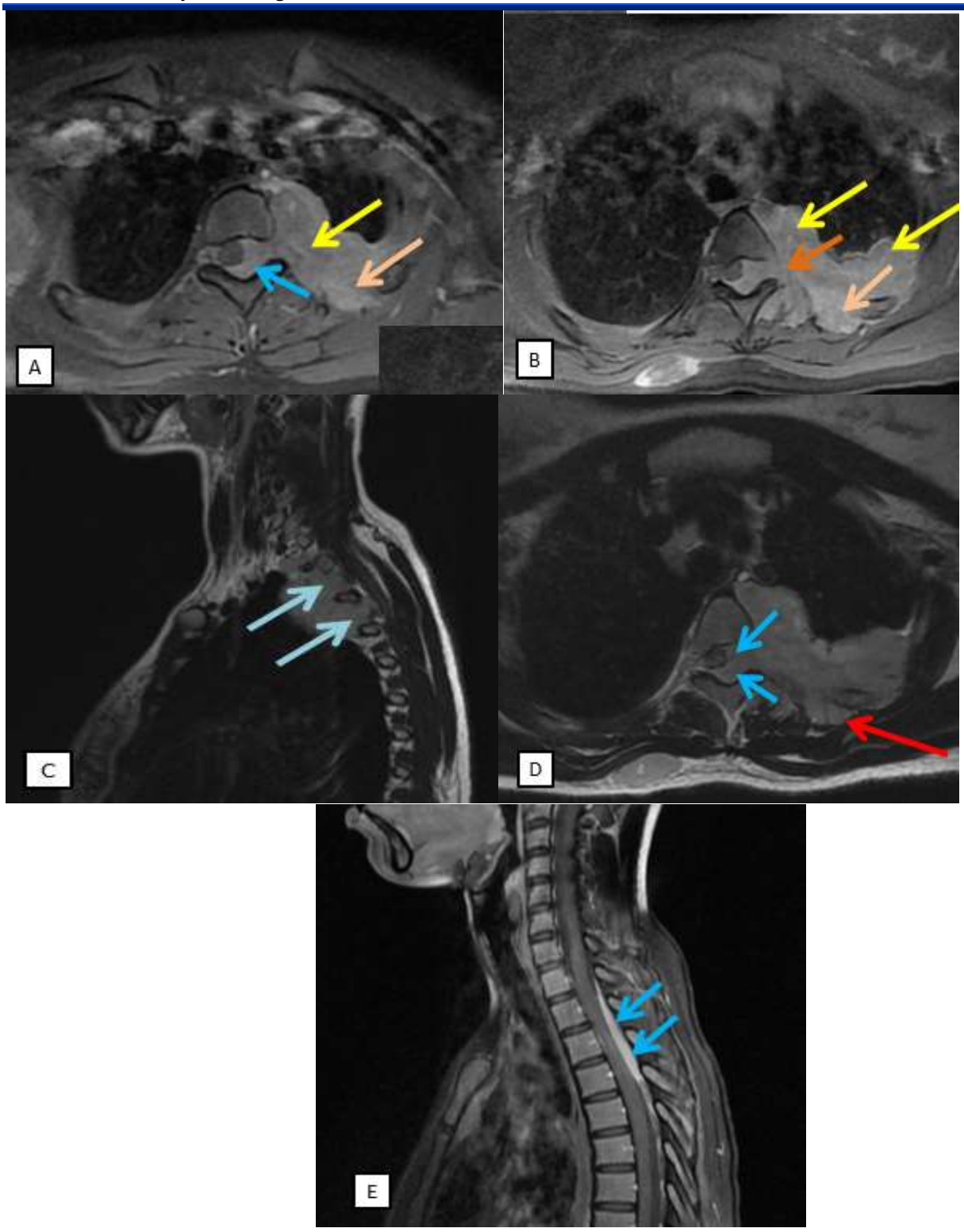

Figure 2: Axial sections through the thoracic level, in T1 FAT SAT sequence, after gadolinium injection (A, B), sagittal (C) and axial (D) T2, and sagittal T1 after injection (E). Left apical pleural thickening, mamelinated, reaching 30 mm in thickness, invading the posterior arch of the first 3 ribs , with extension to the intercostal space and to the adjacent dorsal soft tissues . This thickening also presents an intra-canal invasion at the D1, D2 and D3 levels. It extends to the epidural space, pushing back the medulla , with reduction of its surface. It also presents a foraminal involvement with invasion of the roots.

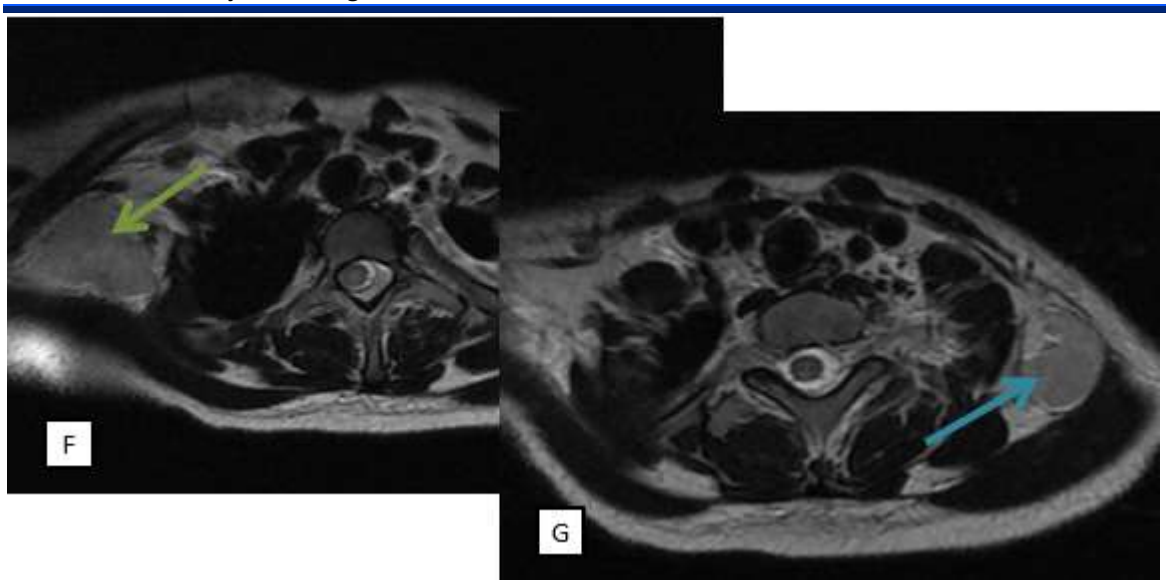

Figure 3: Axial sections through the thoracic stage in T2 sequence (F, G). Presence of a magma of supraclavicular adenopathies bilaterally, measuring 34 x 43 mm on the right and 27 x 20 mm on the left .

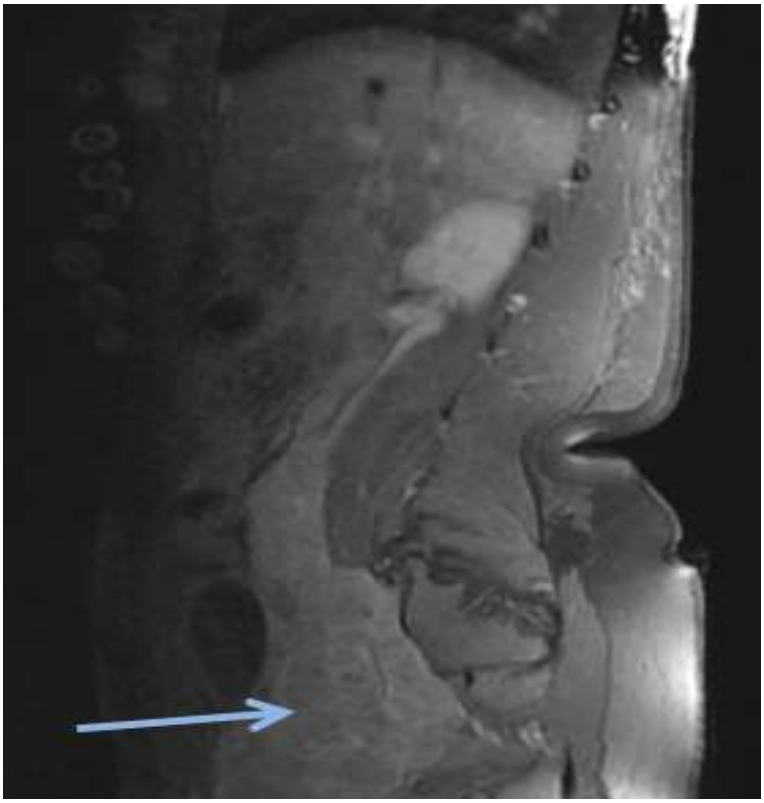

Figure 4: Sagittal sections through the abdomino-pelvic region, in T1 injected sequence (H). Retroperitoneal and right pelvic lymph nodes along the iliac vessels.

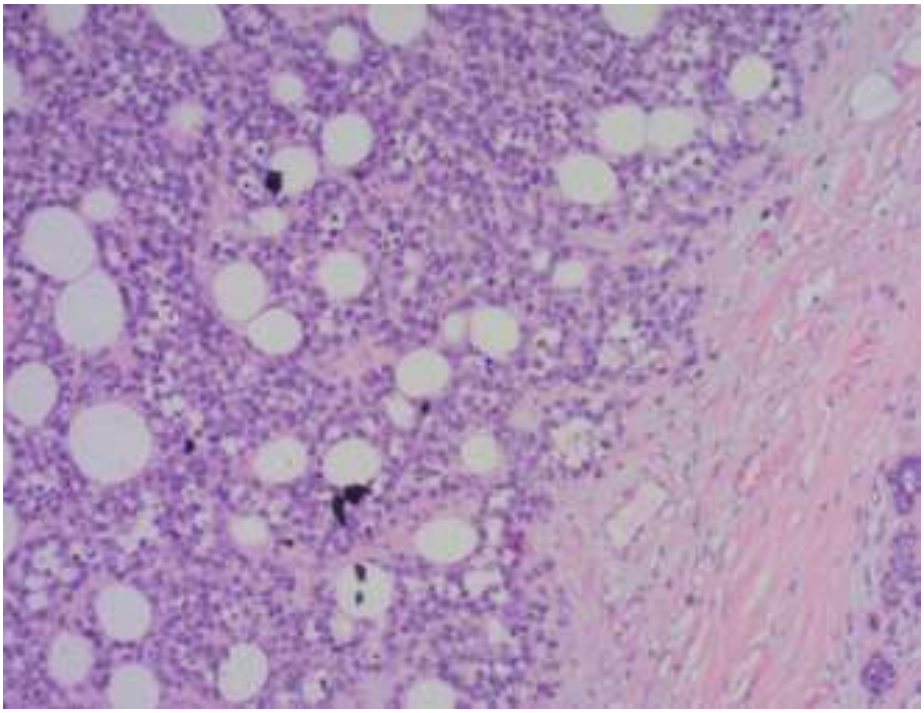

Figure 5: Histologic tumor features showing pseudoalveolar collections of atypical mesenchymal cells with sparse cytoplasm. Hematoxylin and eosin staining

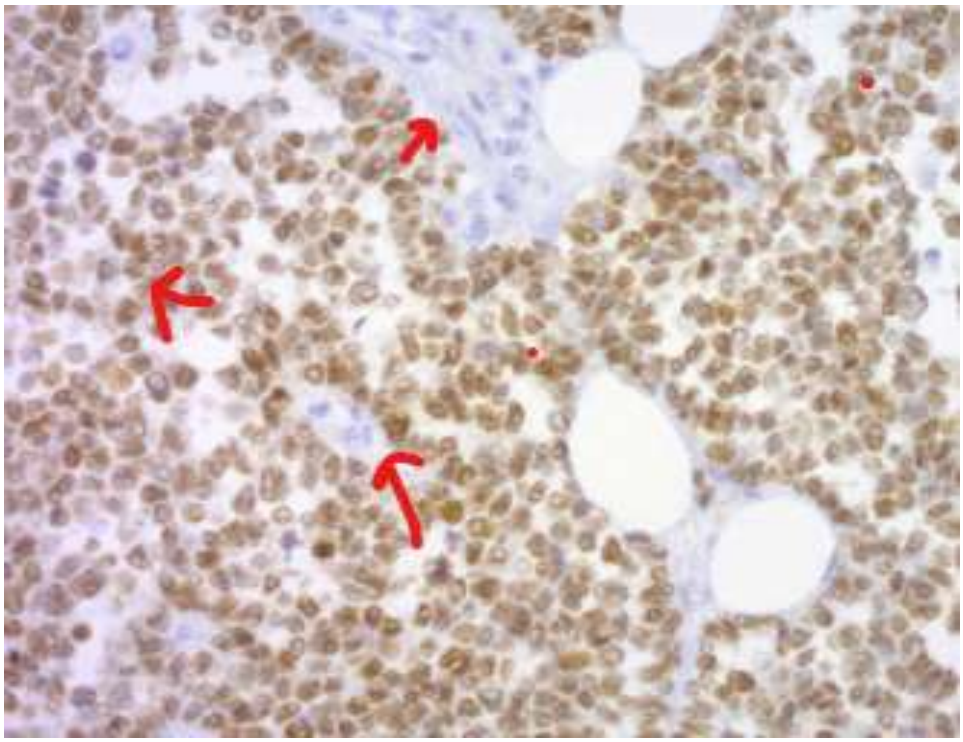

Figure 6: Immunohistochemical staining showing anti myogenin Acpositive tumor cells

Conflict of interest:

The authors declare no conflict of interest.

Figures :

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Références :

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