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Multifocal Ewing Sarcoma with Pancreatic and Cutaneous Metastasis-A Rare Case Report

Case Report

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Abstract

We present a rare case of Multifocal Ewing sarcoma with pancreatic and cutaneous metastasis in a 13-year-old boy who initially presented with unprovoked epistaxis and symptoms of chronic rhinosinusitis. Distinguishing Ewing sarcoma from similar sinonasal tumours is challenging through clinical and radiological assessments alone, necessitating precise histopathological, immunohistochemical, and cytogenetic analyses. The involvement of multiple bones at the time of diagnosis is unusual, and the presence of cutaneous and pancreatic metastasis is exceptionally rare. These unique features pose diagnostic complexities, highlighting the necessity of a comprehensive, multidisciplinary approach for both diagnosis and management.

Keywords: Ewings Sarcoma; Cutaneous Metastasis; Pancreatic Metastasis; Histopathology; Immunohistochemistry

Introduction

Ewing sarcoma is a malignant tumor that can develop in either bones or soft tissues and has the potential to arise in various parts of the body, although it primarily manifests in bones. Initially coined as "diffuse hemangio-endothelioma of bone" by James Ewing in 1921, he theorised that it originated from the endothelial cells lining the blood vessels within the bone. However, the precise cell of origin remains a subject of ongoing debate [1]. This cancer is highly aggressive, especially in children, and ranks as the second most prevalent primary bone tumor. While it can affect nearly any bone, the trunk and long bones are the most frequently impacted.

Typically, Ewing sarcoma presents as a single bony abnormality. In rare instances, it may involve multiple bones at the time of diagnosis. In this paper, we report an unusual case of Multifocal Ewing sarcoma presenting with bilateral scapular lesions, para-nasopharyngeal mass lesion, pancreatic and cutaneous lesions. These combinations of clinical findings are a very rare entity and, as far as we know, no similar case has been documented in the English literature.

Case Report

A 13-year-old boy presented with history of frequent mild to moderate unprovoked epistaxis from left nostril in the past 7 days. His other complaints were intermittent nasal blockage and rhinorrhea and pain in both legs. There was also history of weight loss and loss of appetite. No fever, easy fatiguability, easy bruisability or blood transfusion. Clinical examination showed a polyp-like mass with crusting in left nostril and a 4 x 4 cm mass in oral cavity on the left side in the posterior pharynx. His systemic examination was normal. Basic hematological and biochemistry profile was normal.

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As the patient is a13-year-old male with a nasal mass with unprovoked epistaxis, the clinical diagnosis of juvenile nasopharyngeal angiofibroma (JNA) was considered and the differential diagnoses included, inverted papilloma and sinonasal carcinoma, which werecommoner for the unilateral sinonasal mass. CECT Neck and PNS was advised for further evaluation.

CECT neck and paranasal sinusesshowed a large heterogeneously enhancing mass lesion involving the left para-nasopharyngeal, masticator and pharyngeal mucosal spaces bulging into the nasopharynx. The lesion did not show arterial phase enhancement as would be expected in JNA, showed heterogeneous enhancement in venous phase, involving the muscles of the masticator space (Figure 1). There was an enlarged peripherally enhancing necrotic left level II lymph node as well (Figure 2). Considering age and the above features, the imaging diagnosis of a small round cell tumor, likely rhabdomyosarcoma was proposed.Inverted papilloma is a benign tumour and would not have metastatic deposits, hence was ruled out. The differentials included sinonasal carcinoma. Metastasis was also given as a differential as the lower CT cuts at the thoracic inlet showed a heterogeneously enhancing lesion in the visualised portions of left scapula with adjacent abundant soft tissue. CECT thorax was advised for complete evaluation of the scapular mass.

Meanwhile biopsy was done from the oral cavity mass as the possibility of JNA was ruled out. The biopsy was suggestive of poorly differentiated round cell malignancy (Figure-3). Given the various differential diagnosis of small round blue cell tumors in nasopharynx, an immunohistochemical test was carried out. To our surprise neoplastic cells were positive for CD99, NKX2.2 and negative for CK, LCA, P63, CK5/6, CD56, desmin, chromogranin, myogenin and INSM1 (Figure 4) and (Figure 5). FISH for EWSR1 gene was sent and was positive and confirmed the diagnosis of Ewing Sarcoma.

Further, CECT thorax, was done whichshowed multiple skeletal lesions with large soft tissue components exhibiting aggressive periosteal reaction involving bilateral scapulae, multiple ribs and vertebral bodies. (Figure 6) and (Figure7). Enhancing subcutaneous nodule in the left anterior chest wall was seen (Figure 8) and heterogeneously enhancing pancreatic lesions involving the head, uncinate process and tail regions were also noted (Figure 9)and (Figure 10). Ultrasound images of the subcutaneous nodule in the right anterior chest wall and the pancreatic tail region are depicted

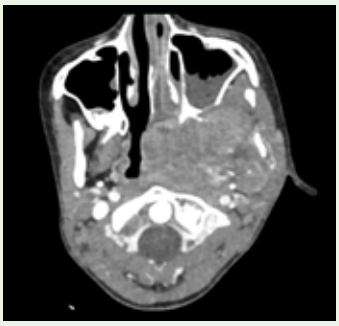

Figure 1: Axial CECT section in soft tissue window showing a heterogeneously enhancing mass lesion involving the left para-nasopharyngeal, masticator and pharyngeal mucosal spaces bulging into the nasopharynx.

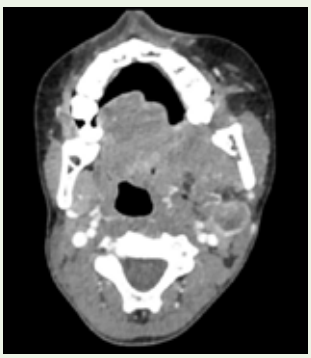

Figure 2: Axial CECT section in soft tissue window showing an enlarged peripherally enhancing necrotic left level II lymph node.

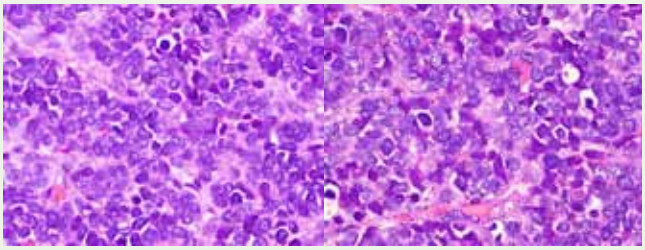

Figure 3: Microscopic images showing sheets and nests of small round blue cells displaying marked nuclear atypia with stippled chromatin, scant eosinophilic cytoplasm and mitotic figures (40X).

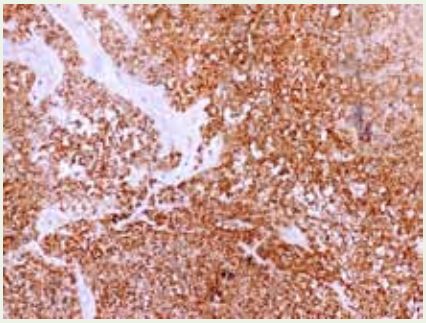

Figure 4: IHC for CD99 showing strong diffuse membranous positivity in the tumor cells.

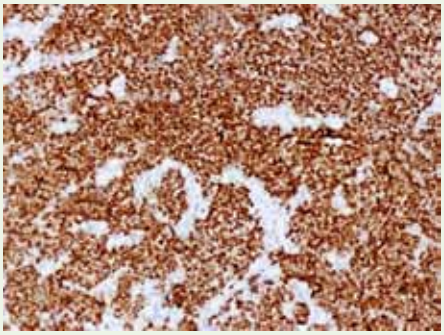

Figure 5: Photomicrograph showing nuclear immunoreactivity for NKX2.2

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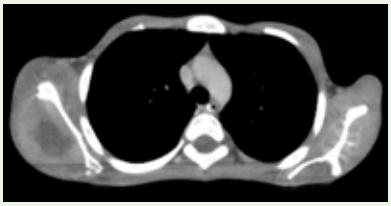

Figure 6: Axial CECT section in soft tissue window showing heterogeneously enhancing soft tissue on either side of bilateral scapulae with aggressive type of periosteal reaction.

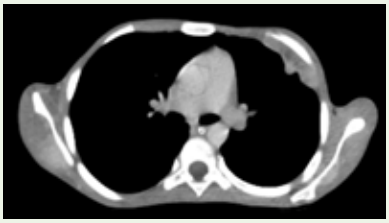

Figure 7: Axial CECT section in soft tissue window showing heterogeneously enhancing soft tissue component arising from the left anterior 3rd rib.

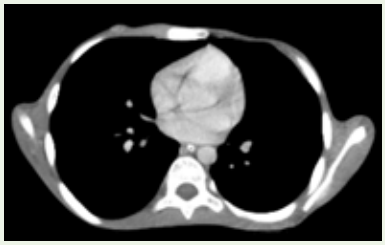

Figure 8: Axial CECT section in soft tissue window showing heterogeneously enhancing soft tissue density lesion in the subcutaneous plane in the right anterior chest wall.

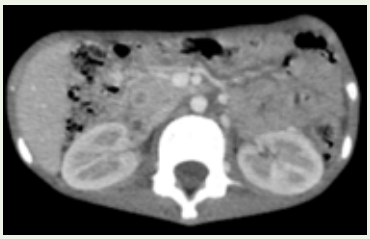

Figure 9: Axial CECT section in soft tissue window showing peripherally enhancing lesion in the head of the pancreas.

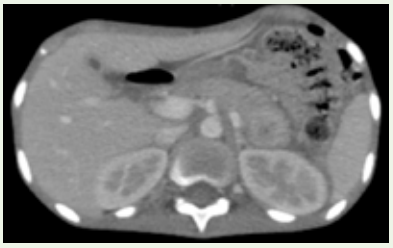

Figure 10: Axial CECT section in soft tissue window showing another heterogeneously enhancing lesion in the tail of the pancreas.

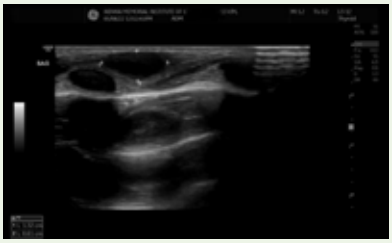

Figure 11: Ultrasound B scan image in sagittal plane using linear probe showing hypoechoic subcutaneous lesion measuring 1.3 x 0.6 cm seen separately from the underlying rib.

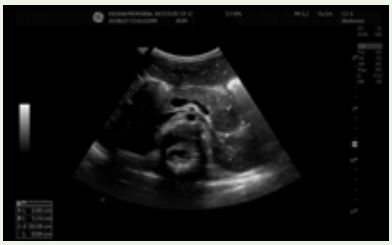

Figure 12: Ultrasound B scan image in axial plane using curvilinear probe showing round to oval hypoechoic lesion in the tail of pancreas.

in (Figure11) and (Figure 12). FNAC from the subcutaneous nodule and the pancreatic lesion was done under ultrasound guidance which showed malignant small round blue cells suggesting cutaneous and pancreatic metastasis.

Bone marrow biopsy was done to rule out marrow involvementand showed intertrabecular spaces replaced by infiltrating neoplasm displaying similar morphology. Patient was started on standard chemotherapy with VAC (vincristine, adriamycin, and cyclophosphamide) alternating with IE (ifosfamide and etoposide). Patient showed stable response after 3 cycles of chemotherapy.

Discussion

Ewing sarcoma, peripheral primitive neuroectodermal tumor (PNET), and Askin tumor are grouped as a single entity due to their common genetic characteristics. These tumors are now collectively referred to as the Ewing sarcoma family and are distinguished by distinct genetic fusions involving FUS, EWSR1, TAF15 (FET), and E26-specific (ETS) FET-ETS genes [2,3]. While these tumors typically arise in bones, they can also manifest in the nasal and sinus regions, often causing vague symptoms. Ewing sarcoma in the head and neck is rare, accounting for only 1-4% of cases, and it's even rarer in the nasal and sinus areas [4,5].

In this case, as the patient presented with symptoms of frequent mild to moderate unprovoked epistaxis and features of chronic rhinosinusitis, our initial differential diagnosis included juvenile angiofibroma, sinonasal carcinoma andinverted papilloma which were more commoner for the unilateral sinonasal mass.

However,after the imaging and biopsy investigations,we were convinced we were dealing with something else.

Distinguishing Ewing sarcoma from similar sinonasal

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tumours based solely on clinical and radiological assessments is challenging. Therefore, an accurate diagnosis typically requires histopathological, immunohistochemical and cytogenetic analyses. Immunohistochemistry usually shows strong CD99 expression while lacking markers associated with muscles and blood cells. In this instance, CD99 and NKX2.2 were positive, while other markers were negative, confirming the diagnosis of Ewing sarcoma.

The imaging finding that hints towards the diagnosis is the bone centric appearance of the lesions with exuberant soft tissue on either sides of bilateral scapulae with aggressive periosteal reaction and not much of cortical destruction. Ewing sarcomas are usually found in the shafts of long bones and tend to extend into nearby soft tissues. Bilateral scapular involvement in a case of Ewing sarcoma as seen in this case is quite unusual.

Cutaneous metastasis is rarely reported in literature [6]. The lesions appear as solitary dermal nodules and they show predilection for young individuals. In our case patient had cutaneous nodules over right side of anterior chest wall and FNAC of which was suggestive of neoplastic small round blue cells.

Our case highlights the rarity of Ewings Sarcoma metastasis to the pancreas as well and till now only 4 cases of Ewings Sarcoma are reported with pancreatic metastasis [7]. It is to be noted that there is no evidence that ESFT is associated with any familial predisposition syndrome or environmental factors [8].

Overall survival rates vary based on whether the cancer has spread at the time of diagnosis. Patients with localized disease tend to have a better prognosis than those with metastatic disease. Prognostic factors include the patient's age, cancer stage, tumour size, and its location. Patients under 15 years of age without metastasis generally have a more favourable outcome compared to those with metastasis.

In summary, Ewing sarcoma occurring in the head and neck is a rare occurrence, and its accurate diagnosis depends on histopathology and immunohistochemistry. Prognosis is significantly influenced by factors such as the presence of metastasis and the patient's age.

Conclusion

Ewing sarcoma presents a wide histological variety, leading to complex diagnostic issues that demand expertise in immunohistochemistry and cytogenetics. Ewing sarcoma in the head and neck is infrequent, and its presence in the sinonasal region is even more rare. Our case highlights the rare presentation of Ewings Sarcoma with cutaneous and pancreatic metastasis making the diagnosis more challenging. It is a multimodality approach which is required for the diagnosis and management for Ewing sarcoma, which involves various investigations. An effective treatment plan including chemotherapy and surgery or chemoradiotherapy is needed for local remission and healing with a better survival rate.

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The patient's guardian (Mother) consent was taken for the study.

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