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Adamantinoma Like Ewing's Sarcoma of Maxillary Sinus: A Rare Entity

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Abstract

Adamantinoma-like Ewing sarcoma (ALES) is a rare variant of the Ewing sarcoma family characterized by epithelial differentiation in a tumor harboring molecular features of conventional Ewing sarcoma. From a pathological standpoint, it poses a significant diagnostic challenge due to its overlapping morphology with basaloid carcinomas and other small round blue cell tumors. Histologically, ALES demonstrates nests and sheets of small round to basaloid cells with peripheral palisading, rosette-like structures, and occasional squamoid differentiation. Immunohistochemically, the tumor exhibits a distinctive biphenotypic profile with diffuse CD99 expression along with cytokeratin and p40/p63 positivity. The presence of EWSR1 gene rearrangement or other FET-ETS fusions confirms the diagnosis. Accurate recognition is essential, as misinterpretation may lead to inappropriate management strategies.

Keywords: ***Adamantinoma; Ewing sarcoma; Maxillary sinus***

1. Introduction

Adamantinoma-like Ewing sarcoma represents a unique morphologic variant of Ewing sarcoma characterized by epithelial differentiation, thereby creating a diagnostic overlap between mesenchymal and epithelial malignancies. It most commonly arises in the head and neck region, particularly the sinonasal tract and salivary glands, where it mimics a wide range of basaloid neoplasms. Histologically, the tumor displays nested or lobular architecture composed of monomorphic small round cells within a fibrotic stroma, often with peripheral palisading and focal keratinization. Despite these epithelial features, ALES is defined by underlying molecular alterations, most commonly EWSR1::FLI1 fusion, placing it within the Ewing sarcoma spectrum. Recognition of this entity requires careful integration of morphology, immunohistochemistry, and molecular findings [1-3].

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2. Case Report

A 47 years old male presented with complaints of right sided nasal bleed since 8 months and right sided cheek swelling since last 2 months. On CECT neck, relatively well defined heterogenous soft tissue mass epicentered in the right maxillary sinus with minimal enhancement measuring 4.5 cm × 4.2 cm × 5.1 cm. Anteromedially the mass was extending into right nasal cavity, premaxillary subcutaneous tissue and medially causing nasal septum deviation and postero-laterally causing extension into masticator space.

Then, the patient was evaluated further evaluated and biopsy from both nasal cavity and oral cavity was sent for histopathological examination. After processing the biopsy and staining with Hematoxylin and Eosin (H&E), the slides were examined under light microscopy. At low power, multiple necrotic and ulcerated bits of tissue were identified along with some areas of tumor cells. These tumors cells were arranged in diffuse sheets, nests and arranged around blood vessels. These cells were mildly pleomorphic having high N:C ratio, round to oval nuclei, coarse chromatin, inconspicuous nucleoli and scant cytoplasm. Another population of tumor cells have moderate pleomorphic nuclei, vesicular chromatin and conspicuous nucleoli and moderate eosinophilic cytoplasm. Mitosis was brisk and areas of necrosis was seen. Histopathology it was reported as poorly differentiated malignant tumor (FIG. 1 a-b).

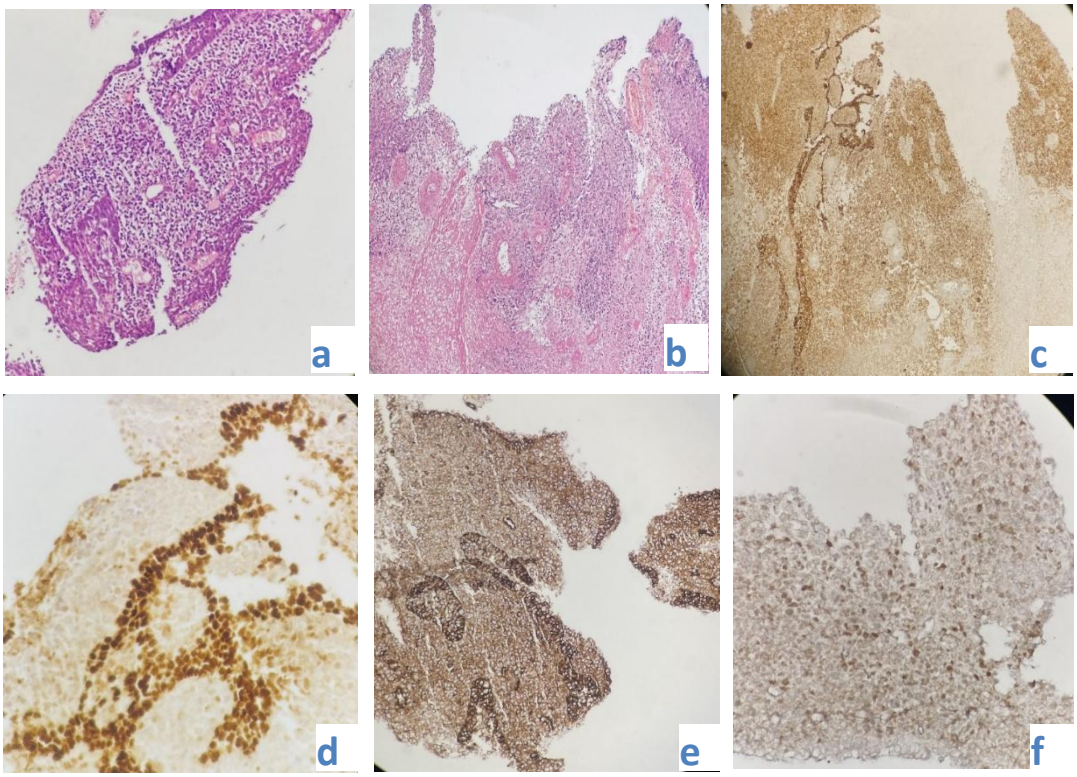

FIG. 1. **(a), (b)- Photomicrograph showing tumor cells arranged in diffuse sheets having high N:C ratio with hyperchromatic nuceli and scant cytoplasm along with (b) some areas showing perivascular arrangement of tumor cells (200X, H&E). (c), (d), (e) and (f) Photomicrograph showing Immunohistochemical positive staining for PanCK (diffuse cytoplasmic), p40 (diffuse nuclear), CD 99 (diffuse membranous) and NKX 2.2 (Diffuse nuclear Positive), 200X Magnification.**

On performing Immunohistochemistry (IHC), the epithelial components of tumor cells were diffuse positive for PanCK, p63 and p40. On further evaluating, CD99 was diffuse membranous positive and NKX2.2 was nuclear positive in malignant round cells (FIG. 1 c-f). Proliferative Index was very high (90%-95%) and Vimentin was diffuse cytoplasmic positive. These tumor cells were negative for CD45, CD3, CD20, S-100, EBV-LMP1, Synaptophysin, EMA, CK7, IDH, CD117, CD56, Chromogranin, CK5/6, CK20, myogenin and desmin. Further molecular evaluation was needed but due to financial constraints, it was not done. Thus, a final diagnosis of Adamantinoma like Ewing's Sarcoma was made.

3. Discussion

From a histopathological perspective, ALES demonstrates a heterogeneous growth pattern comprising solid nests, sheets, and cords of small round to basaloid cells embedded in a fibrous or hyalinized stroma. Tumor cells typically exhibit scant cytoplasm, hyperchromatic nuclei, and inconspicuous nucleoli. A characteristic feature is peripheral palisading, imparting a basaloid appearance, along with pseudoglandular or rosette-like structures. Foci of squamous differentiation and keratinization may also be present, further contributing to its resemblance to epithelial malignancies specially basaloid neoplasms of sino-nasal tract likely basaloid squamous cell carcinoma, NUT carcinoma, sino-nasal undifferentiated carcinoma, olfactory neuroblastoma etc [3,4].

Immunohistochemically, ALES shows a distinctive dual phenotype. There is strong, diffuse membranous expression of CD99 along with nuclear FLI1 and NKX2.2 positivity, supporting Ewing sarcoma lineage. Concurrently, epithelial markers such as pancytokeratin (AE1/AE3), CK5/6, and p40/p63 are expressed, sometimes diffusely. Neuroendocrine markers like synaptophysin may show focal positivity, whereas markers such as S100, desmin, and LCA are typically negative. This biphenotypic immunoprofile is a key diagnostic feature but can also lead to confusion if interpreted without molecular correlation [1,3].

The differential diagnosis is broad and includes basaloid squamous cell carcinoma, olfactory neuroblastoma, sinonasal undifferentiated carcinoma, and NUT carcinoma, all of which may share overlapping morphologic features. However, the presence of diffuse CD99 positivity and demonstration of EWSR1 rearrangement helps distinguish ALES from these entities. Conventional Ewing sarcoma may also be considered but typically lacks strong epithelial marker expression. Importantly, recent studies have also identified alternative fusions such as FUS::FEV in rare cases, emphasizing the need for extended molecular testing when EWSR1 rearrangement is not detected [4,5].

4. Conclusion

Adamantinoma-like Ewing sarcoma is a rare and diagnostically challenging tumor that requires a high index of suspicion in cases showing basaloid morphology with aberrant epithelial differentiation. The coexistence of CD99 positivity and cytokeratin expression, along with molecular confirmation of EWSR1 or related gene rearrangements, is essential for definitive diagnosis. Accurate pathological identification is critical, as it directly influences therapeutic decisions, which follow Ewing sarcoma treatment protocols rather than carcinoma-based approaches. Increased awareness and the use of comprehensive diagnostic modalities are vital to avoid misdiagnosis and ensure optimal patient management [1,2,5].

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