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Adamantinoma like Ewing sarcoma of pelvis- A rare tumor at an uncommon site

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Abstract
Adamantinoma like Ewing Sarcoma is a distinct, easily identifiable clinico-pathologic entity with distinct morphologic and molecular features. It's a rare variety of the Ewing family of cancers that mostly affects the head and neck area. Lack of follow-up data, including treatment and outcome, limits our ability to fully understand the natural history of this disease.

Keywords: Ewing's sarcoma, adamantinoma like variant (ALES), pelvis

Introduction
Ewing's sarcoma is a highly malignant tumor of bone and soft tissue. James Ewing, who initially identified it as a separate entity, is honored in its name. The second-most frequent primary malignant bone tumor in children is Ewing Sarcoma, which has a distinct preference for those between the ages of 10 and 20. Although the precise cause is unknown, it has been shown that a distinctive t (11; 22) chromosomal translocation is present in 85% of cases [1]. The pelvis and lower extremity are where Ewing sarcoma is most frequently found, Iliac bone is the origin in 12.5% of cases. In a clinical setting, pain predominates as a presenting symptom in 90% of instances, followed by swelling in 70% of cases. There are several different types of Ewing sarcomas with varied histological characteristics, including the classic, atypical/large cell, sclerosing, spindle cell, and one with squamous differentiation [2].
The word "adamantinoma" refers to a tumour that histologically resembles ameloblastomas of the jaw. It is a rare form of Ewing sarcoma that accounts for 0.1%–0.5% of cases and is characterized for having histological and immune-histochemical signs of squamous differentiation. Although the relocation of the skin's basal epithelium during embryological development is the most widely accepted notion, the etiology of ALES is still a topic of debate [3, 4]. Van Haelst published the first description of a rare Ewing Sarcoma variation that shared morphological similarities with Adamantinoma in 1974. He classified the patient as either a Ewing sarcoma with squamoid differentiation or a poorly differentiated Adamantinoma [5]. Due to the presence of a EWSR1 gene rearrangement, it was later genetically confirmed to be a Ewing sarcoma with epithelial differentiation and was given the name adamantinoma like Ewing sarcoma (ALES).
Full understanding of the natural history of ALES is somewhat limited by a paucity of follow-up information, with treatment and outcome data. We discuss one such case in which a patient with discomfort, tenderness, and swelling in the right iliac fossa in which later Possibility of Ewing family of tumour, adamantinomatous variant was suggested.

Case Report
A 21-year-old female, Asian Indian in origin presented to the surgery OPD with history of swelling which was asymptomatic for over a year and then suddenly progressed along with pain in the right iliac fossa. Pain was moderate in intensity. There was no history of trauma, known infection or contact with tuberculosis. There was no involvement of any other joints.No rectal and urinary complaints were mentioned.
On examination, there was localized tenderness over the right sacroiliac joint. Lumbar spine and hip examination were normal.

The left sacroiliac joint was also normal. The peripheral joints and neurological examination were normal as well. Investigations revealed an erythrocyte sedimentation rate of 45 mm, haemoglobin of 10.8 g/dL and total leucocyte count of 12,500. The serum chemistry and urine analysis was normal.

The radiographs demonstrated multiple, confluent lytic regions with sclerotic borders in the supra-acetabular region of right ilium, without significant cortical breech or soft tissue component. MRI with contrast showed an expansile mass lesion arising from posterior aspect of right iliac bone, pubic bone and anterior pillar of acetabulum causing marrow infiltration, surrounding cortical disruption with periosteal reaction and significant large circumferential soft tissue showing extra and intra pelvic component. Contiguous extension into right hip joint causing erosion of articular surface and antero-medial margin of right femur bone was also noted. Extra pelvic soft tissue component infiltrating right gluteus muscle and extending into right upper thigh involving medial and anterior compartment. Findings were suggestive of neoplastic mass and further biopsy and histopathological correlation was suggested.

Section studied shows tumour arranged in nests, sheets and cords. Tumour cells exhibit basaloid appearance, minimal pleomorphism, hyperchromatic nuclei with scanty cytoplasm (Figure 1 A, B, C). Tumour cells were positive for CK, NSE, FLI1 (Weak, focal), P63, CD99 (Weak, focal) and negative for GFAP, NKX2.2, SATB2, SYNAPTOPHYSIN (Figure 2A, 2B). Final impression of Possibility of Ewing family of tumour, Adamantinomatous variant was suggested. The patient was treated with multidrug chemotherapy using drugs, which include vincristine, doxorubicin, ifosfamide and etoposide.

Discussion

ALES is a unique, recognizable clinic-pathologic entity with specific morphologic, immune-histochemical, and molecular features. ALES often develops in bone or nearby soft tissue. It was originally described in the extremities and thorax, this tumour type has since been predominantly recognized in the head and neck. ALES harbours a recurrent t (11; 22) *EWSR1- FLI1* translocation [6]. Folpe *et al*. [7], in 2005 described unusual variants of Ewing Sarcoma including ALES. He demonstrated the complex immunoprofile of ALES; for instance, unlike in classic Ewing Sarcoma HMWCK was expressed in all the cases of ALES. While the age spectrum of ALES is wide (Range 7–77 years), most (> 80%) patients are younger than 50 years of age (Mean: 36.7 years). Symptoms may be non-specific; most patients present with an enlarging mass, with or without pain. Exceptionally, metastatic disease may be the first presenting site. In present case, patient had swelling for over a year which increased in size along with aggravating pain [8].

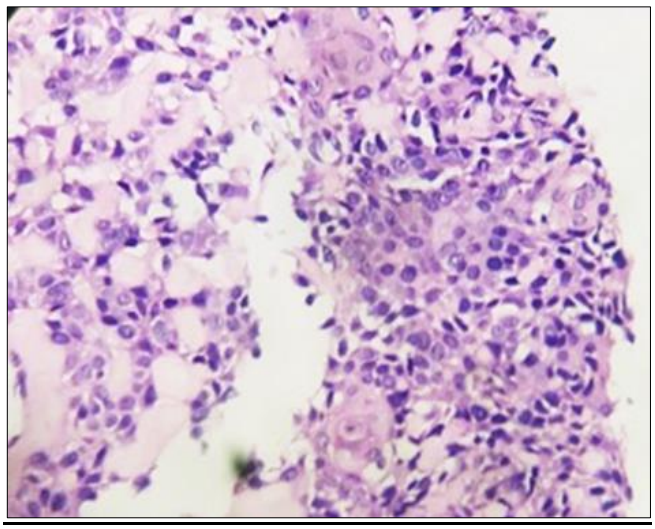
ALES generally presents as a large tumour, with a mean size of 4.3 cm (Range 2.3 to 7.9 cm). While these tumours consistently have infiltrative borders, the extent of invasion differs somewhat by anatomic sites. ALES on radiography frequently demonstrates extensive involvement and destruction of surrounding structures. Upon gross examination, ALES tends to have a white to grey, firm, fibrotic, and lobulated cut surface with variable amounts of cystic degeneration and calcifications [8, 9].

The existence of a basaloid neoplasm with cytologic homogeneity in spite of high grade histologic characteristics including necrosis and an accelerated mitotic rate is the most useful findings for diagnosing ALES. Histologically, ALES is distinguished by epithelial-differentiated small, globular blue cells that frequently resemble clusters of epithelioid/basaloid cells with peripheral nuclear palisading and desmoplastic stroma. Frank squamous differentiation with keratinization can be seen focally. Peripheral nuclear palisading and rosette development are seen in some tumours, however not all tumours exhibit these characteristics. Likewise, somewhat abrupt formation of small, compact keratin pearls is only seen in a minority of cases [10].

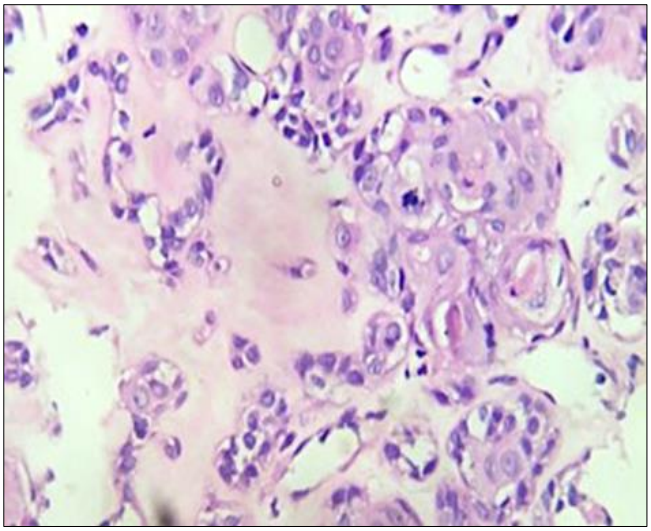
ALES shares the strong, membranous CD99 expression and nuclear NKX2.2 positivity characteristic of conventional Ewing sarcoma. But, by definition, this variant also demonstrates positivity for cytokeratin, p63 and p40. ALES also shows variable and usually focal positivity for neuroendocrine markers. Additionally, almost all cases of ALES have been negative for S100, SMA, desmin, WT1, and NUT1. However, unlike typical ES, a nested growth pattern, peripheral palisading, and a complex epithelial differentiation (Evinced by low- and high-molecular- weight cytokeratins, and p40/p63 immunoreactivity, with or without overt squamous differentiation, or keratin pearls) bestow ALES a deceptive resemblance to carcinomas. These hybrid pathologic features of ES and carcinoma make ALES a distinctive and treacherous entity at the same time. One of the distinctive features of diagnostic importance, identified in about a third of ALES cases is the presence of overt squamous differentiation [11, 12].

Surgical resection, followed by radiation therapy and adjuvant chemotherapy are the available treatment options and outcomes seems to vary according to anatomic sites. Rooper *et al.* [10] in their study reported that of 12 reported cases all of them underwent surgery among which 2 patients treated with VDC/IE (22%) and 2 patients treated with other regimens (40%). Of all cases at 6 months follow up, 6 patients (50%) experienced persistent, recurrent, or metastatic disease and at last follow up, 8 of these patients (66%) had no evidence of disease, 3 patients (25%) were alive with disease, and 1 patient (9%) was dead with disease.

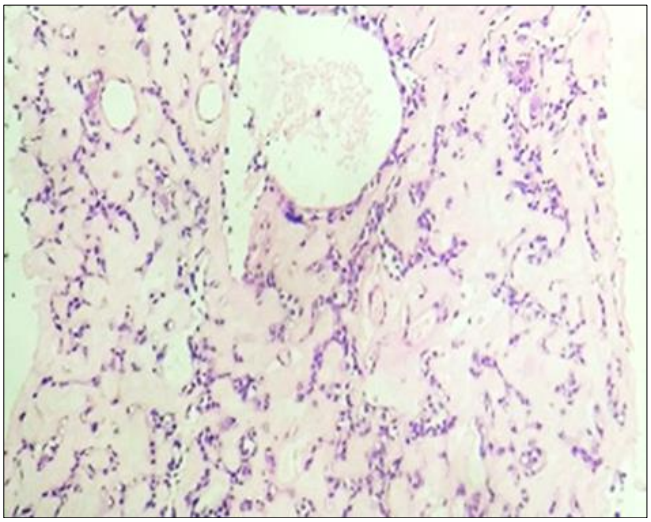
Till now, only a handful of cases of Adamantinoma like Ewing family tumour have been reported; most of them are in the head and neck region (Sinonasal tract, thyroid, salivary glands etc.). Our case is a rare occurrence of Adamantinoma like Ewing family tumour in the pelvis.



A



B



C

Fig 1: A, B, C show tumour arranged in nests, sheets and cords with basaloid appearance and minimal pleomorphism

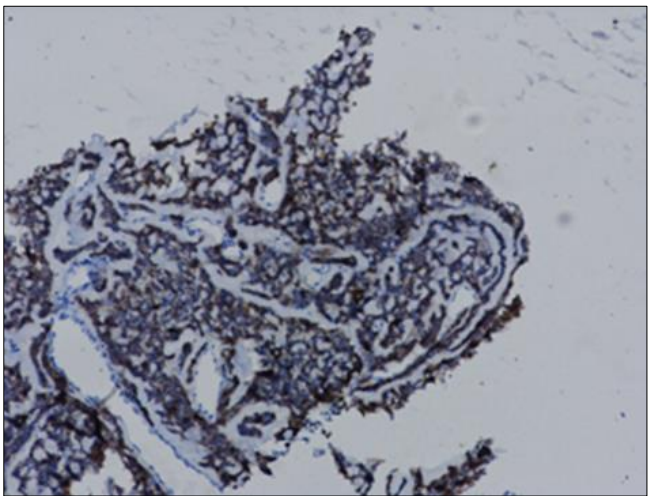


Fig 2A: Positive for PanCK (AE1/AE3)

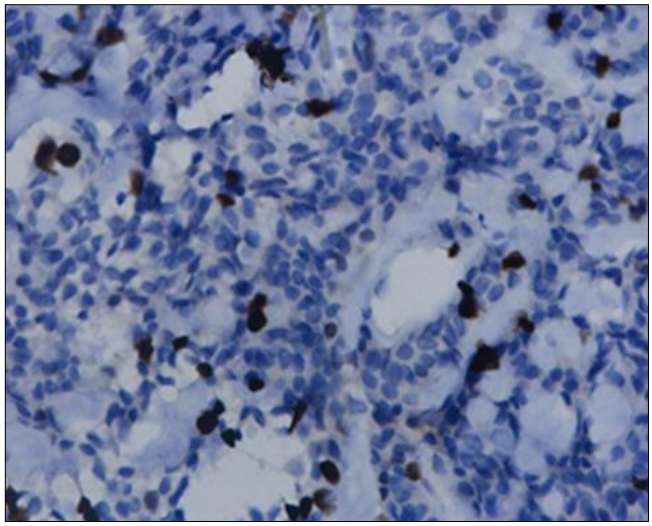


Fig 2B: Ki-67-25%

Conclusion

To sum up, Adamantinoma, like Ewing Sarcoma, is a rare variation of the Ewing family of tumours that primarily affect the bone and nearby soft tissue of the extremities as well as the head and neck region. These tumours also exhibit variable epithelial differentiation and a complex immunological profile, making them susceptible to misdiagnosis. To definitively identify this rare, aggressive variation of the Ewing family that exhibits epithelial differentiation, CD 99 and NKX2.2 should be included in the evaluation of poorly differentiated and undifferentiated tumors that arise in children and young adults. To precisely identify and subclassify Adamantinoma like EFT, molecular studies for EWSR1 gene rearrangement should be conducted after positivity for pertinent immunohistochemical markers demonstrating both components.

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