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Epithelioid Sarcoma

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Summary:

A 22 years old male patient complaining of erythematous ill-defined multinodular firm mass with areas of ulceration affecting his left thumb & the thenar eminence of 3 years duration. Histopathological examination revealed hyperplastic epidermis with multiple dermal nodules composed of epithelioid cells with eosinophilic & pleomorphic cytoplasm. Immuno-histochemical studies revealed that the cells are positive for vimentin, pancytokeratin and epithelial membrane antigen (EMA).

Introduction:

Epithelioid sarcoma (ES) was first described by Enzinger in 1970. It's a sarcoma simulating a granuloma or a carcinoma. This rare malignant tumour occurs in adolescents and young adults. It is usually located on the distal extremities (hands, fingers and forearm); rarely occurs on the pelvis, perineum and vulva [1].

Case presentation:

A 22 years old male patient presented to the outpatient clinic at Al-Haud Al-Marsoud Hospital, complaining of erythematous ill-defined multinodular firm mass with areas of ulceration affecting his left thumb and the thenar eminence of 3 years duration, with history of recurrence after previous excision **(fig. 1)**.

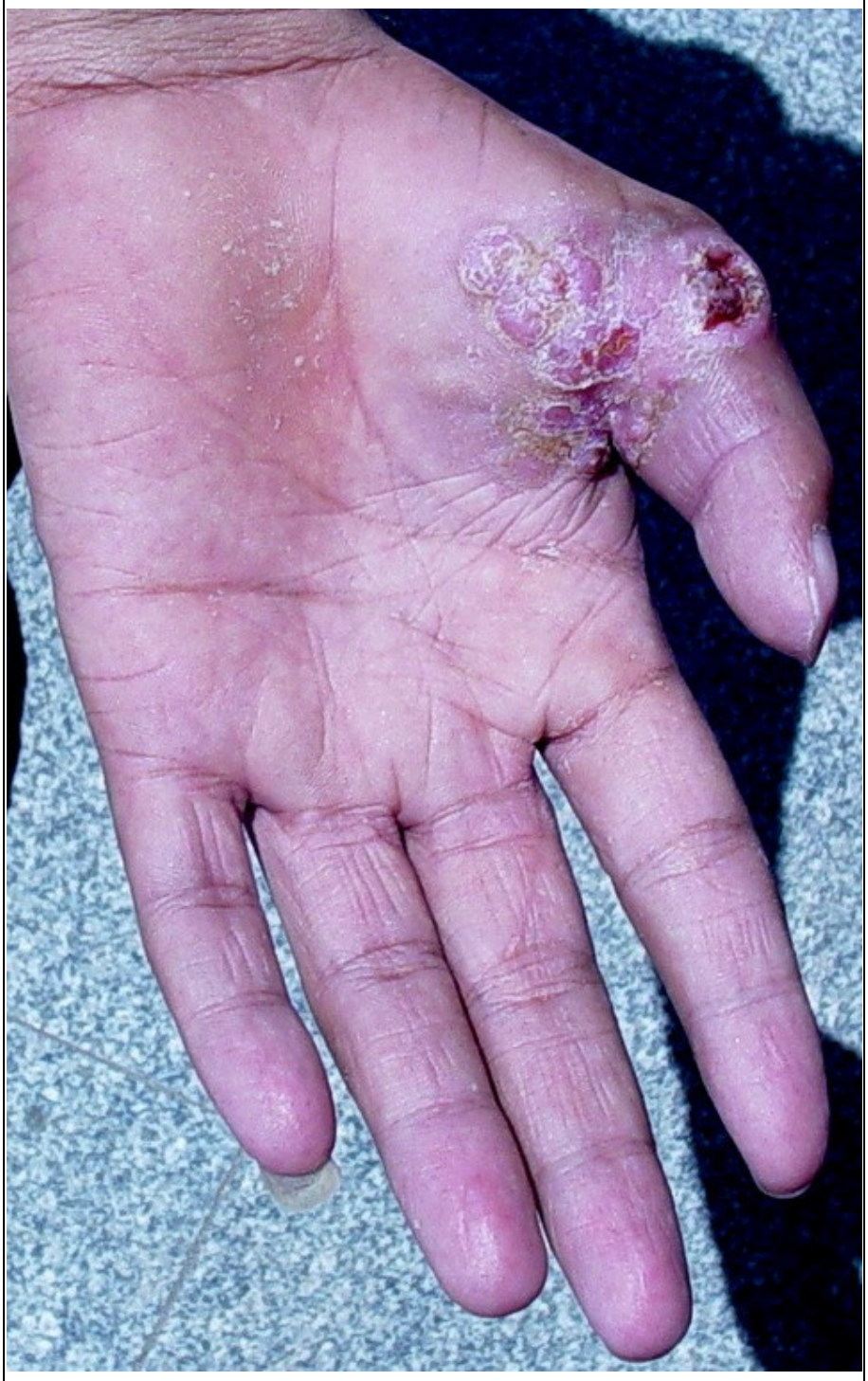

Fig 1: Erythematous ill defined multi-nodular firm mass with areas of ulceration affecting the left thumb and the thenar eminence.

Our clinical differential diagnoses of the case included: atypical mycobacterial infection, lupus vulgaris, squamous cell carcinoma, low grade malignant vascular tumor and angiosarcoma.

Histopathological examination revealed hyperplastic epidermis with multiple dermal nodules composed of epithelioid cells with eosinophilic and pleomorphic cytoplasm. The nodules were embedded in collagenous fibrous stroma, and surrounded by lymphocytic infiltrate with areas of haemorrhage and necrosis. Higher power examination showed the tumour cells surrounding central zones of necrosis (**fig. 2- 6**). So according to these findings, we excluded atypical mycobacterial infection, lupus vulgaris and squamous cell carcinoma.

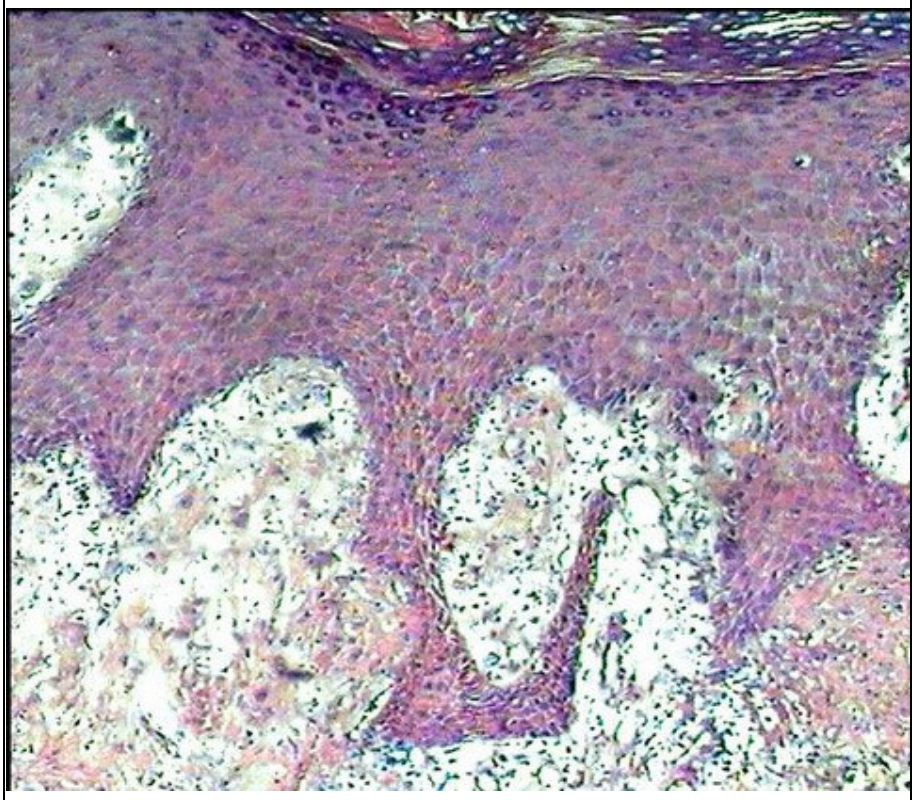

Fig 2: H&E stained section showing hyperplastic epidermis.

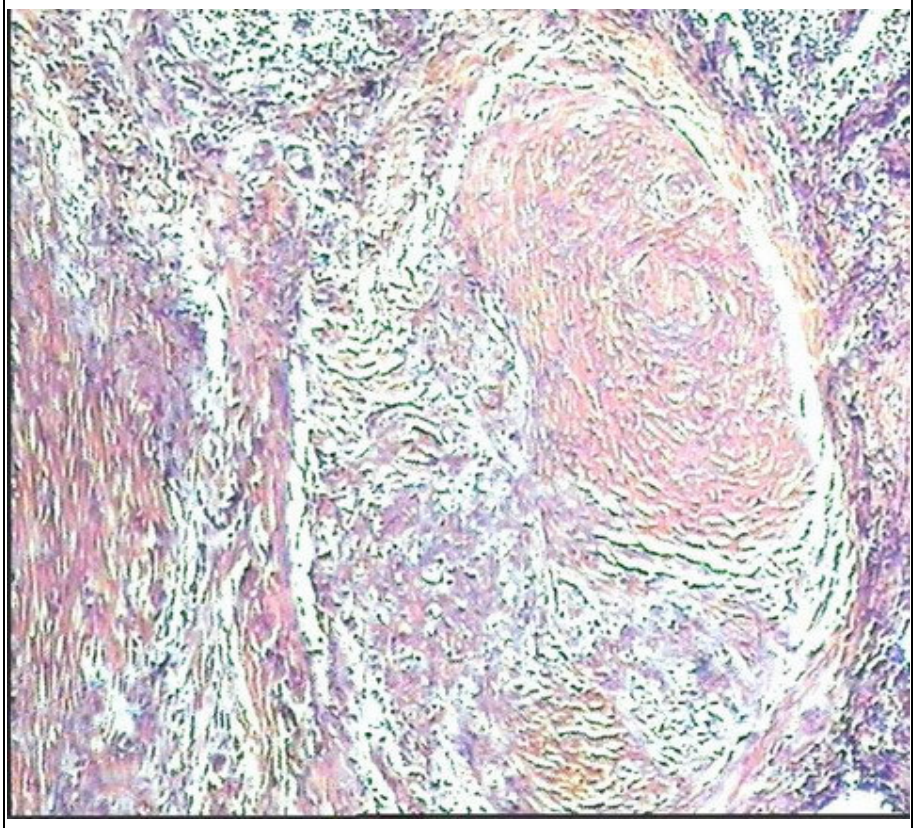

Fig 3: H&E stained section showing multiple dermal nodules embedded in collagenous fibrous stroma.

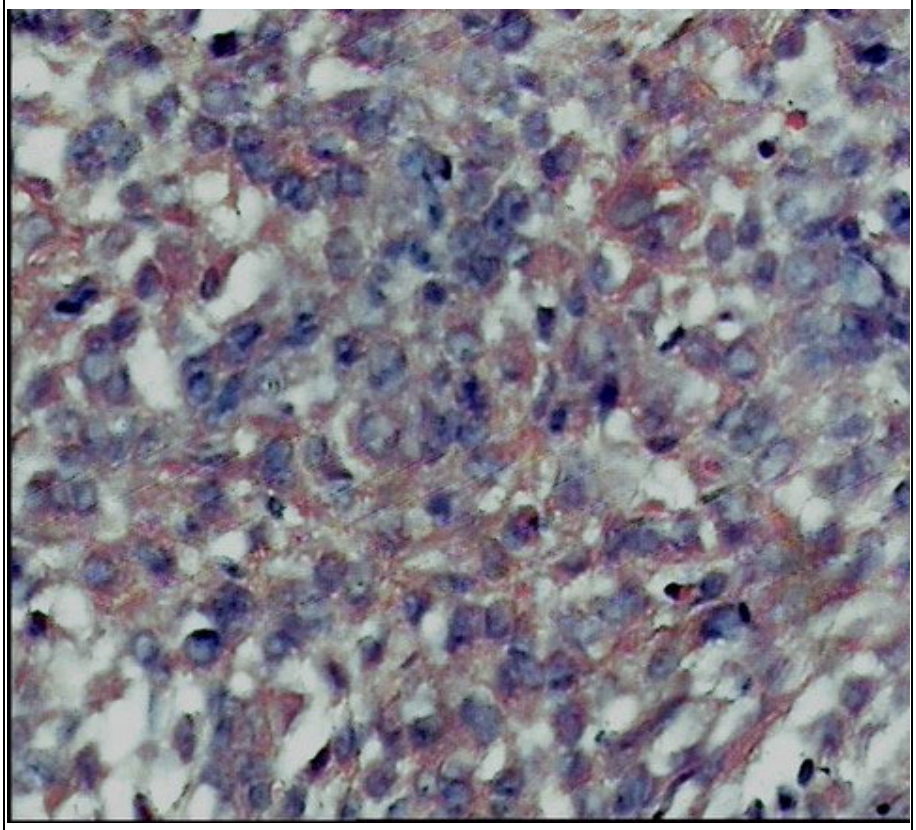

Fig 4: The nodules are composed of epithelioid cells with eosinophilic & pleomorphic cytoplasm.

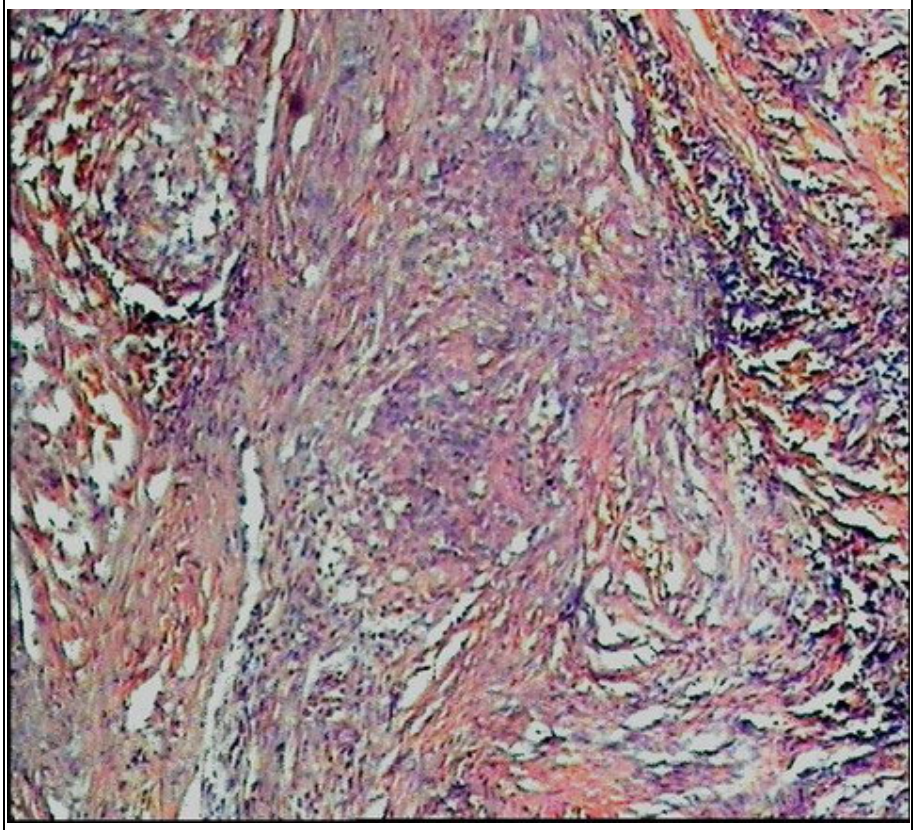

Fig 5: Nodules are surrounded by lymphocytic infiltrate, areas of haemorrhage and necrosis.

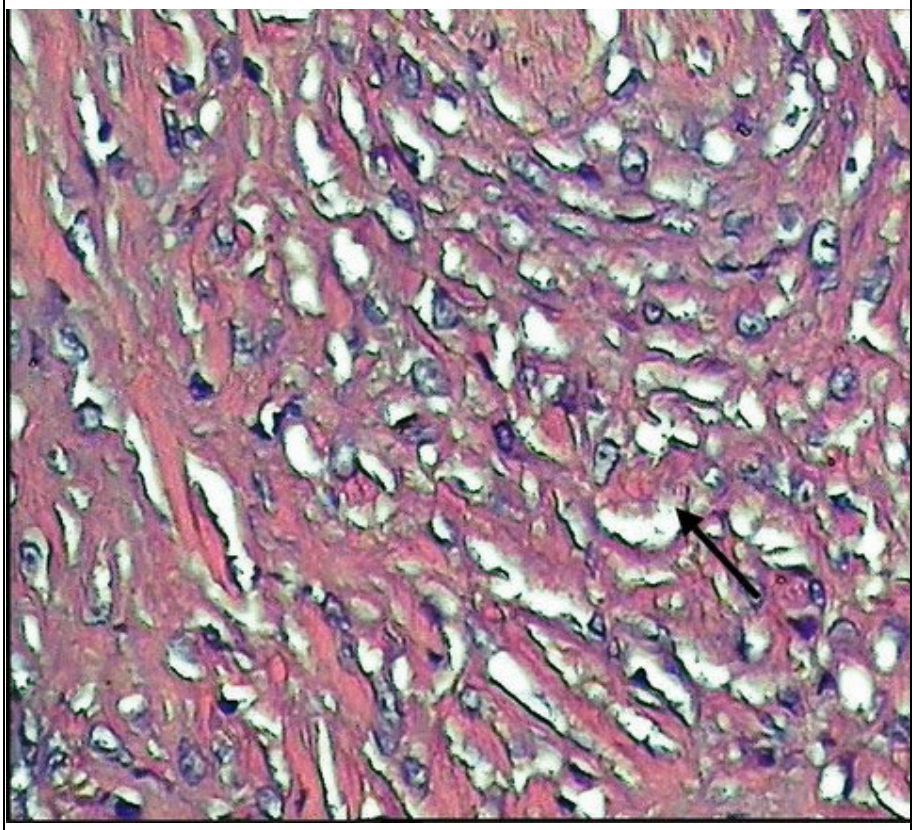

Fig 6: Tumor cells surrounded central zones of necrosis.

Immuno-histochemical studies were done for further assessment of vascular origin of the tumour, but surprisingly the cells were negative for CD 34 **(fig. 7)**, factor 8 **(fig. 8)** and S100 protein **(fig. 9)**, leading us to exclude these vascular entities (low grade malignant vascular tumor and angiosarcoma). So, we proceeded with other immune-histochemical markers which revealed that the cells are positive for EMA **(fig. 10)**, pancytokeratin **(fig. 11)** and vimentin **(fig. 12)**, which are consistent with epithelioid sarcoma which was our final diagnosis.

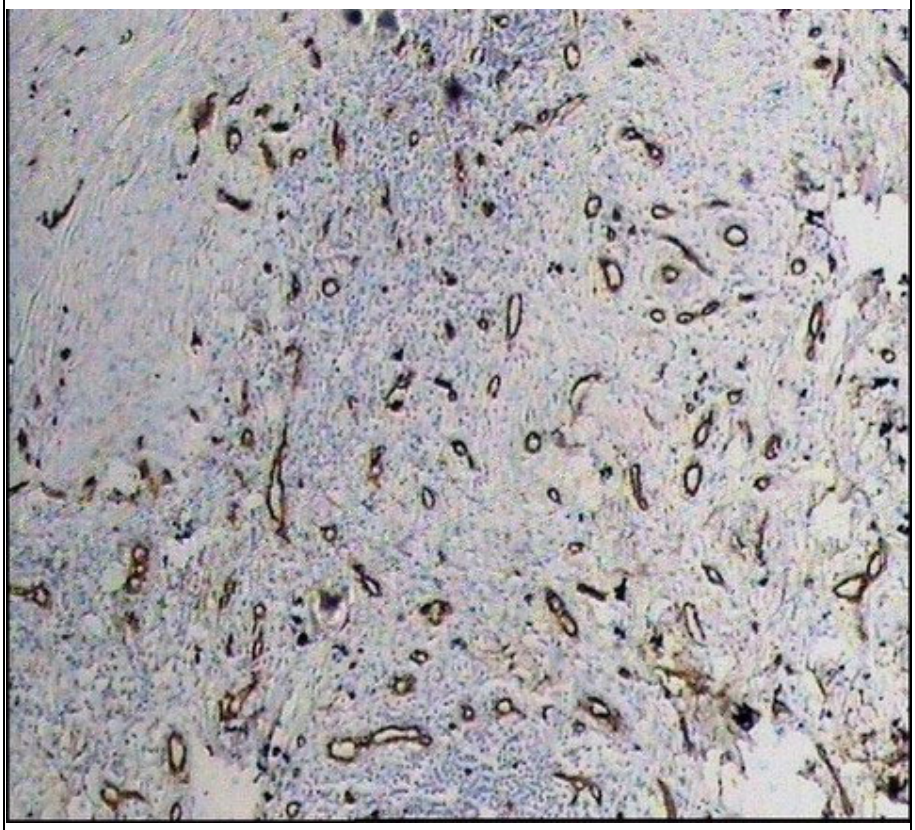

Fig 7: CD34 was negative.

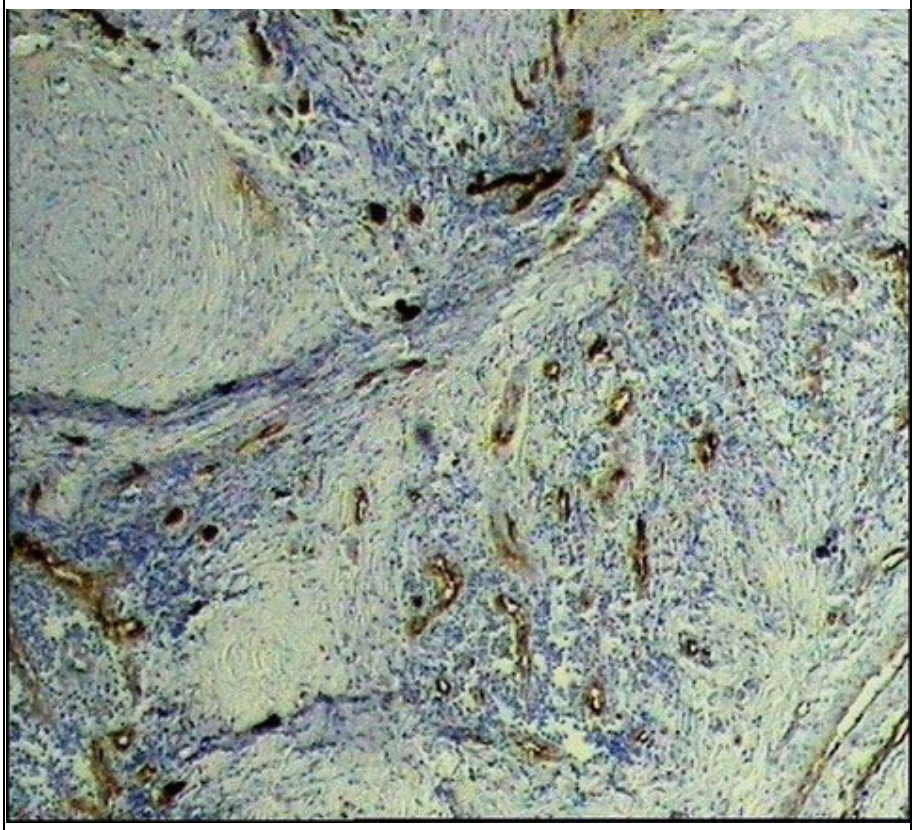

Fig 8: Factor 8 was negative.

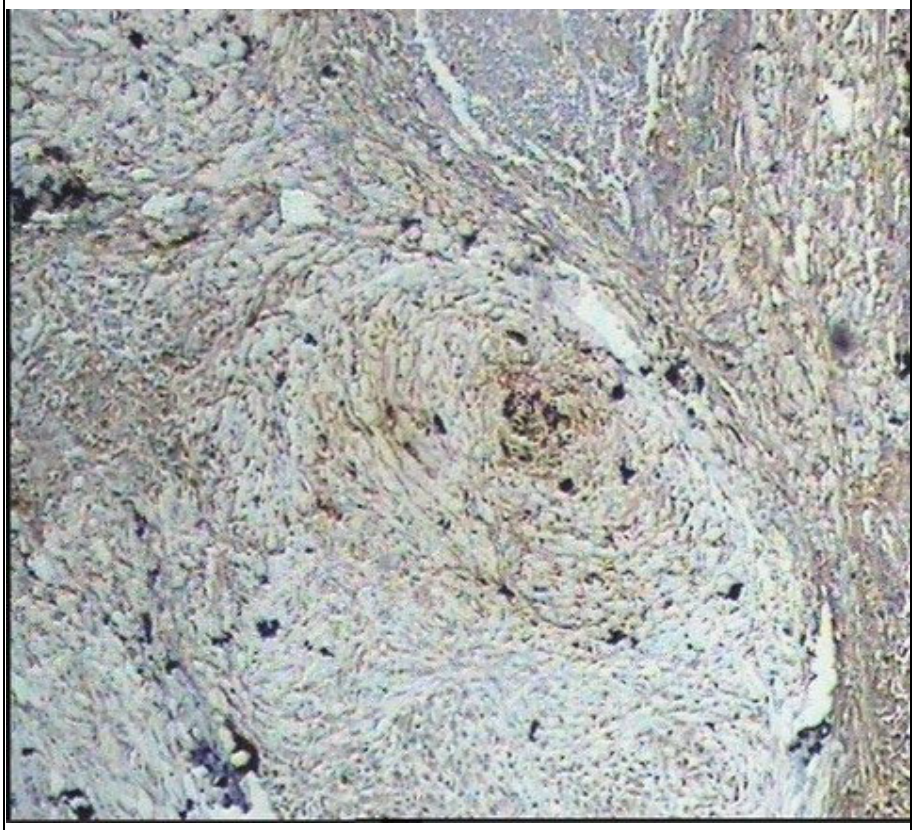

Fig 9: S100 protein was negative.

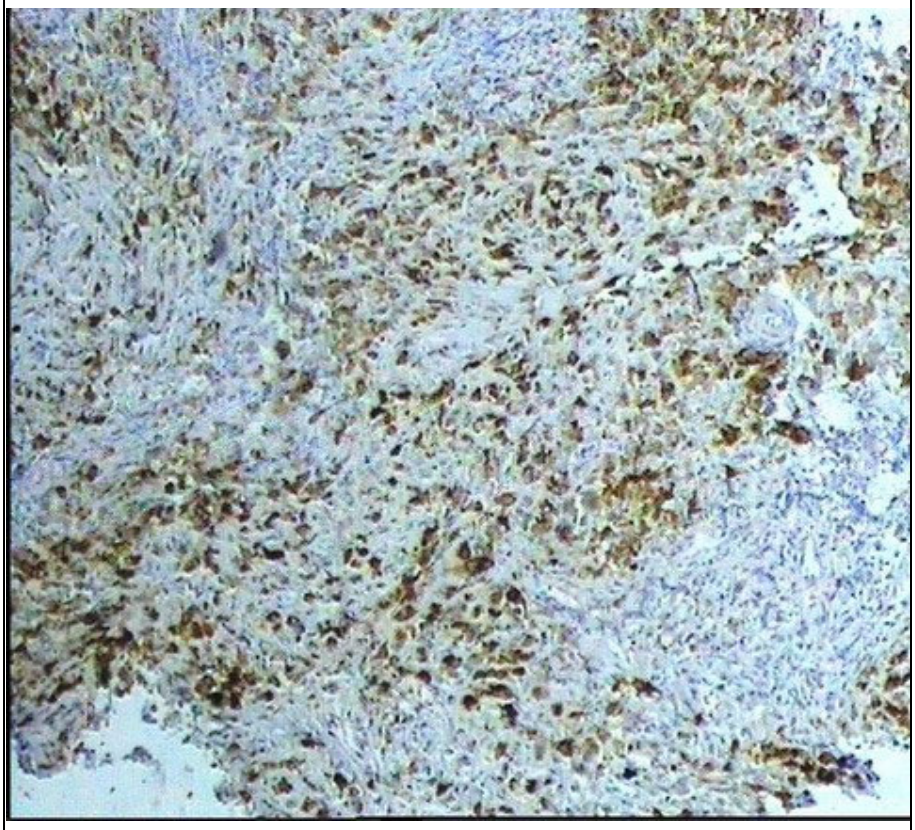

Fig 10: EMA was positive.

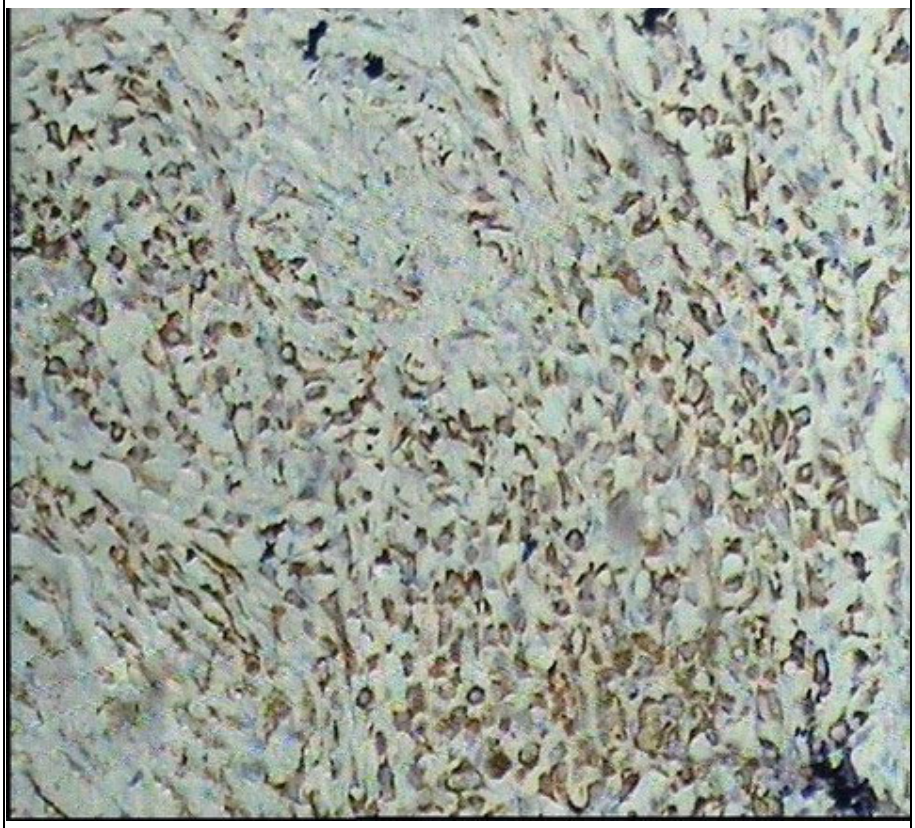

Fig 11: Pan- cytokeratin was positive.

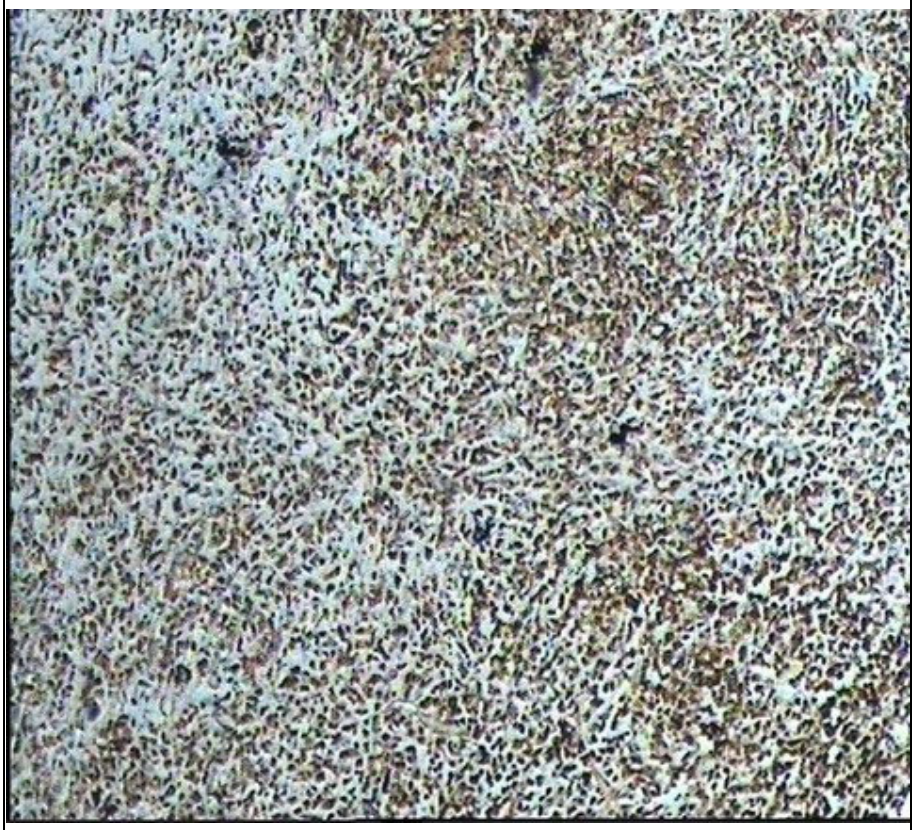

Fig 12: Vimentin was positive.

Discussion:

Epithelioid sarcoma is a distinctive malignant soft-tissue tumour composed of cells with epithelial differentiation. It is an uncommon tumour, affecting males more often than females and tending to begin in early adult life [1,2]. The presenting sign can be a painless slowly growing firm, dermal nodule or lobular subcutaneous tumour that grows outwards and may ulcerate early. These nodules surrounded by fibrous tissue and fat. The cut surface is grayish-white and flecked or mottled with yellow or brown spots, reflecting the presence of areas of necrosis. The distal extremities are the usual situation for the tumour, particularly the flexor aspect of the finger and the palm [3,4].

Microscopically the nodules are composed masses of large, round, polygonal or spindle cells with acidophilic cytoplasm. The larger nodules have necrotic centers and show the so-called 'geographical necrosis', which may be mistaken on scanning power microscopy for a granuloma. Tumour cells show clear histological, ultrastructural and immune-histochemical evidence of epithelial differentiation. Local recurrence after excision is common and metastasis, principally to lymph nodes, lung and pleura may occur [5,6].

Immuno-histochemically, tumour cells of ES are positive for keratin and EMA. They also express vimentin, and in 50% of the cases they are positive for CD34 [7].

Complete removal by surgical excision is essential if local recurrence and eventual metastasis are to be avoided. Surgical excision followed by radiotherapy is often recommended. Involvement of regional lymph nodes is associated with distant metastasis and death. The survival rate has been estimated to be between 65% and 70% [5,8]. Features associated with poorer prognosis include male sex, older age at diagnosis, proximal location, tumour size, mitotic rate, necrosis, vascular invasion and local recurrence and lymph node metastasis [5,6,8].

References

1. Enzinger F. Epithelioid sarcoma: a sarcoma simulating granuloma or carcinoma. Cancer 1970; 26: 1029- 1041

2. Fletcher CDM, McKee PH. Sarcomas: a clinic-pathological guide with particular reference to cutaneous manifestation. Clin Exp Dermatol 1984; 9: 451- 465

3. Santiago H, Feinerman LK, Lattes R. Epithelioid sarcoma: a clinical and pathologic study of 9 cases. Hum Pathol 1972; 3: 1706- 1710

4. Evans HL, Baer SC. Epithelioid sarcoma: a clinic-pathologic and prognostic study of 26 cases. Semin Diagn Pathol 1993; 10: 286- 291

5. Halling AC, Wollan PC, Pritchard DJ, Vlasak R, Nascimento AG. Epithelioid sarcoma: a clinicopathologic review of 55 cases. Mayo Clin Proc 1996; 71: 636- 642

6. Prat J, Woodruff JM, Marcove RC. Epithelioid sarcoma: an analysis of 22 cases indicating the prognostic significance of vascular invasion and regional lymph node metastases. Cancer 1978; 41: 1472- 1487

7. Arber DA, Kandalaft PL, Mehta P, Battifora H. Vimentin-negative epithelioid sarcoma: the value of an immunohistochemical panel that includes CD34. Am J Surg Pathol 1993; 17: 302- 307

8. Chase DR, Enzinger FM. Epithelioid sarcoma: diagnosis, prognostic indicators and treatment. Am J Surg Pathol 1985; 9: 241- 263.