

## Epithelioid sarcoma of the orbit

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Accepted: March 2010

Ann Saudi Med 2011; 31(2): 187-189

PMID: \*\*\*\* DOI: 10.4103/0256-4947.75589

Epithelioid sarcoma is an aggressive and rare malignancy first recognized by Enzinger in 1970. It is known most commonly to affect the distal upper extremities in young adults. The classical “distal” form has a male predominance and can also involve other less frequent sites including lower extremities, proximal upper extremities, and the trunk. The “proximal” variant of this tumor is deep seated, tends to occur in older patients and predominantly develops in the pelvis, perineum, and genital tract. In the orbit, only a single report of two cases, which had a typical histopathologic appearance, has been previously published. We present the third case of orbital primary epithelioid sarcoma.

Epithelioid sarcoma is an aggressive and rare malignancy first recognized to affect the distal upper extremities in young adults. Less frequent sites include the lower extremities, the proximal upper extremities, and trunk.<sup>1,2</sup> The etiology is unknown, but it is postulated to arise in the tendon sheaths with a history of preceding trauma in some cases.<sup>2</sup> Recently a “proximal” variant of epithelioid sarcoma has been identified that predominantly develops in the pelvis, perineum, and genital tract. The tumor in these cases is deep seated and tends to occur in older patients.<sup>3</sup> In the orbit, a single report of two cases, which had a typical histopathologic appearance, was previously published in 1993.<sup>4</sup> We present a third case of orbital primary epithelioid sarcoma.

### CASE

A 5-month-old male healthy infant presented to our hospital with 1 month history of right proptosis. He was a product of a full-term pregnancy with normal development. The examination revealed severe right-side proptosis with marked conjunctival chemosis and prolapse. The right eye was displaced inferiorly by a firm superior orbital tumor. MRI revealed a superior extraconal orbital mass measuring 3.4×2.6 cm. The mass was well-defined with minimal heterogenous enhancement and cystic changes. A right orbital biopsy was performed with the initial impression of lymphangioma.

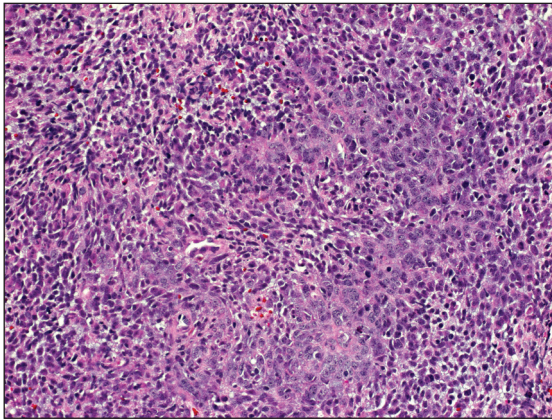
The histopathology showed a multinodular pattern

of growth with large pleomorphic epithelioid cells, abundant cytoplasm, vesicular nuclei, and prominent nucleoli (Figure 1). The tumor nodules showed central necrosis with pseudogranulomatous appearance. Mitotic figures were frequent (Figure 2). There was strong staining for vimentin (Figure 3) and CD34 (Figure 4), in addition to moderate positivity with cytokeratin (Figure 5). Tumor cells were negative to desmin and myogenin.

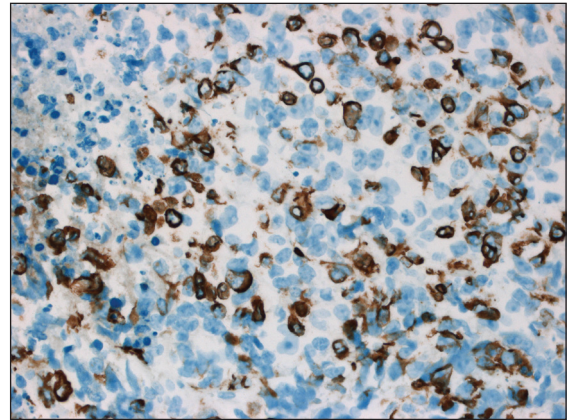
The diagnosis of epithelioid sarcoma “proximal type” was made. The patient had initial debulking of the tumor and adjunctive chemotherapy (three cycles of non-rhabdoid soft tissue sarcoma protocol over 2 months). There was no response of the residual tumor to the chemotherapy for which right exenteration was performed at the age of 7 months. The patient died 2 months later because of intracranial extension of a recurrent tumor.

### DISCUSSION

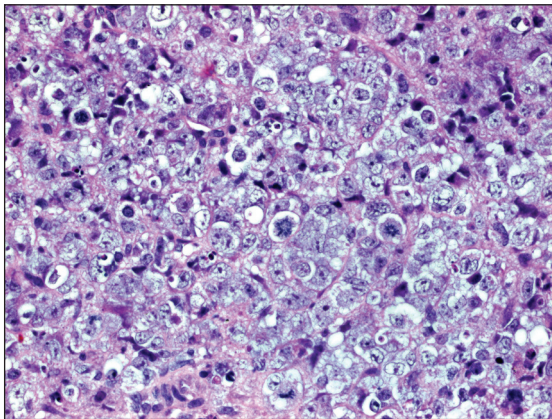
Epithelioid sarcoma is a rare malignant tumor first recognized by Enzinger in 1970, who later on reported a large series of 241 cases from the Armed Forces Institute of Pathology.<sup>1,2</sup> The classical “distal” form occurs in young adults with a male predominance and the most common site is the distal upper extremity. The more recently described “proximal” type is a deep-seated tumor that develops in older adults and predominantly involves the pelvis, perineum, and genital



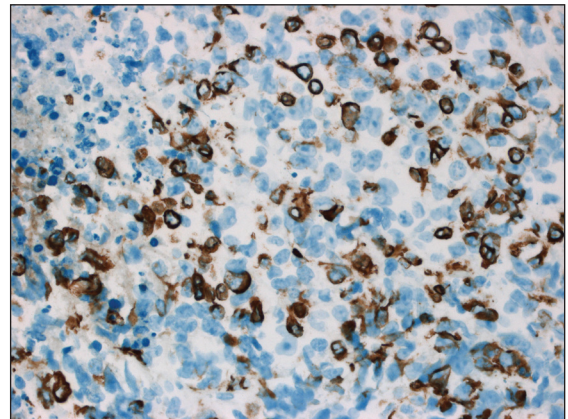
**Figure 1.** Tumor cells with pseudogranulomatous appearance (hematoxylin and eosin,  $\times 200$ ).



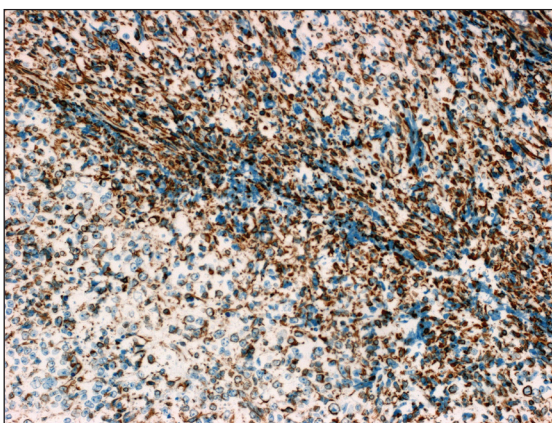
**Figure 4.** Tumor cells with positive staining (CD34,  $\times 200$ )



**Figure 2.** Tumor cells with frequent mitotic figures (hematoxylin and eosin,  $\times 400$ ).



**Figure 5.** Focally positive tumor cells (cytokeratin,  $\times 400$ )



**Figure 3.** Tumor cells with positive staining (vimentin,  $\times 200$ ).

tract.<sup>3</sup> Clinically, most tumors are firm and palpable in the deep soft tissue with symptoms that are related to the site of its development.<sup>1</sup>

The histogenesis is uncertain but there is a possible fibrohistiocytic origin. It is believed by some to arise from mesoderm, which gives rise to the synovial membranes of tendon sheaths and joints.<sup>1,4,5</sup> The head and neck region is seldom involved. The first case of central nervous system primary epithelioid sarcoma was reported in 2002 with involvement of the dura.<sup>6</sup> In the orbit, a single case of metastatic epithelioid sarcoma was included in a series of 51 cases.<sup>7</sup> Two cases of primary orbital epithelioid sarcoma were reported in 1993 involving a 17-year old girl and a 34-year old woman.<sup>4</sup> These were comprehensively presented with detailed histopathologic and immunohistochemical features. The tumor occurred in the supero-temporal and inferior parts of the orbit, which is similar to the location of the tumor in our 5-month old infant. Their histopathologic

description included nodular proliferation of polygonal to spindle-shaped cells with vesicular nuclei and abundant eosinophilic cytoplasm. The tumor had a pseudogranulomatous appearance due to the presence of central necrosis in many of the nodules.<sup>3</sup> Although these two cases were not classified at that time as "proximal", this appearance, in addition to the unique location, would fit into the proximal variant described later in 1997, in which rhabdoid features can also be observed.<sup>3</sup> The immunohistochemical staining profile was identical to our case with a strong vimentin staining, moderate staining for keratin, and negative staining for desmin, S-100, and myoglobin. The second case showed strong staining for an epithelial membrane antigen, but weak staining for vimentin and keratin.<sup>4</sup>

These immunohistochemical properties have been originally described in the classic "distal" form of epithelioid sarcoma.<sup>5,8</sup> The common positivity for CD34 was further realized and added to the profile in 1993, as a helpful stain, especially in vimentin-negative tumors.<sup>9</sup> The unique histopathologic features and the immunohistochemical staining of the tumor in our case have strongly supported our histopathologic diagnosis of epithelioid sarcoma.

The overall prognosis of this tumor is not favorable, with local recurrence in 77% of patients and metastatic disease in 45%, most frequently to lymph nodes and lung.<sup>1</sup> The recurrence depends on the adequacy of the initial excision. Several adverse prognostic indicators have been advocated including male sex, older age, larger tumor size, deep location, and the presence of vascular and/or nerve invasion.<sup>2,7</sup> The most recent study in 2007, including 54 patients, concluded that there is a better prognosis for single localized disease stage absent regional spread to lymph nodes. Their 10-year overall survival was 61.8% with local failure in 26% and distant involvement in 44%.<sup>10</sup> The relentless progression of the disease in the orbit was demonstrated by White et al in their 34-year-old patient who developed two local recurrences following initial piecemeal exenteration. Despite extensive surgical treatment and chemotherapy, the patient died of pelvic metastasis 29 months after her first biopsy.<sup>4</sup> Our patient similarly died of intracranial extension of a recurrent tumor despite surgical intervention and adjunctive chemotherapy. This emphasizes the need for early detection, proper histopathologic diagnosis, and adequate surgical excision of such tumors.

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