Original article

SPINDLE CELL TUMOR OF THE MAXILLA: AN ADULT-TYPE FIBROSARCOMA OR A STORIFORM UNDIFFERENTIATED PLEOMORPHIC SARCOMA?

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ABSTRACT

OBJECTIVE: Spindle cell neoplasms of the jaws include a heterogenous cluster that could pose diagnostic difficulty.

CASE REPORT: A 44-year old woman with left maxillary swelling had histology showing monomorphic fusiform to spindle cells and few mildly pleomorphic cells arranged in a vague herringbone, cartwheel and storiform patterns. Also seen were few foci of epithelioid-like cells and 'staghorn' pattern of vascular channels. Histology differentials included: Fibrosarcoma (adult type), Malignant Peripheral Nerve Sheet Tumor, Hemangiopericytoma, Spindle-cell type angiosarcoma, Synovial sarcoma, Epithelioid sarcoma and Spindle-cell Malignant Melanoma and storiform Undifferentiated Pleomorphic Sarcoma. Immunohistochemistry was done using the antibodies: CD34, CD45, myogenin, synaptophysin, cytokeratin AE1/AE3, desmin, melan-A and vimentin. Vimentin was positive (++) while all others were negative. The final diagnosis was strongly suggestive of intra-osseous adult-type fibrosarcoma, mostly based on morphological features than immunohistochemical distinction.

CONCLUSION: This case highlights one of the difficulties faced in the process of diagnosis by resource-limited facilities. The patient has not presented for any intervention (due to financial challenges) as at the time of this manuscript documentation and submission.

Keyword: Spindle cell tumor, fibrosarcoma, undifferentiated pleomorphic sarcoma

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INTRODUCTION

Spindle cell tumors of the jaws may present significant difficulty for diagnosis, especially with hematoxylin and eosin staining alone. Tumors included in this category are: malignant peripheral nerve sheath tumor (MPNST), undifferentiated pleomorphic sarcoma, fibrosarcoma, epithelioid sarcoma, spindle-cell angiosarcoma, hemangiopericytoma and spindle-cell melanoma.[1] Reaching a final diagnosis, even with ancillary tests like immunohistochemistry, may still be challenging especially because of poor specificity of some immunomarkers for lesions in this category and in low-resource laboratories which may not be

able to accommodate an extensive panel of markers. From the differentials listed above, fibrosarcoma is typically the usual suspect for most spindle cell malignancies of the jaws and the diagnosis of fibrosarcoma is typically by exclusion of close histological mimics.[1]

CASE REPORT

A 44 years old female presented to the Oral Pathology department of University College Hospital, Ibadan with a 5 months history of a painful left maxillary swelling. She also gave history of tooth ache and spontaneous exfoliation of teeth in the same quadrant about a year prior to presentation. Examination revealed a firm, painful mass, measuring approximately 8.0cm X 10.0cm with apparently clinically normal overlying skin. There was associated left-sided epiphora and naso-labial fullness. There was mobility of 22, 23, 24, 25, 26 and 27, and mucosal ulceration and indentations of the lower teeth on the swelling. There was no clinically detectable involvement of cervical lymph nodes.

Radiographs showed osteolysis of the floor, medial wall, lateral wall and part of the roof of the left antrum with associated floating of 24, 25 and 27 (Figure 2a and 2b). Microscopic examination of the biopsy specimen revealed monomorphc cells admixed with a few mildly pleomorphic cells. These cells have moderate eosinophilic cytoplasm and fusiform to spindle shaped nuclei. Some epithelioid-like cells having moderate cytoplasm, hyperchromatic nuclei and prominent nucleoli were also seen. No abnormal or increased mitosis was observed. The spindle cells are disposed in fascicles, storiform, cartwheel and herringbone-like patterns. The epithelioid cells were disposed in sheet and poorly formed nodules. Neoplastic cells are in a well vascularized connective tissue stroma with irregularly shaped blood vessels reminiscent of a staghorn (Figure 3a and 3b). Based on the microscopic examination, the following were considered as possible differentials: fibrosarcoma (adult-type), MPNST, hemangiopericytoma, spindle type angiosarcoma, synovial sarcoma, epithelioid sarcoma, undifferentiated pleomorphic sarcoma and spindle cell melanoma.

Based on these differentials, an immune-panel was selected, based on available resources. This included; vimentin, myogenin, desmin, CD34, S-100, Melan-A, AE1/AE3 and synaptophysin (Figure 4). All markers were negative except vimentin which was positive.

Figure 1. Clinical photograph of the patient showing left maxillary swelling with intra-oral mucosal ulceration. Picture B is the patient on follow-up after 3-months.

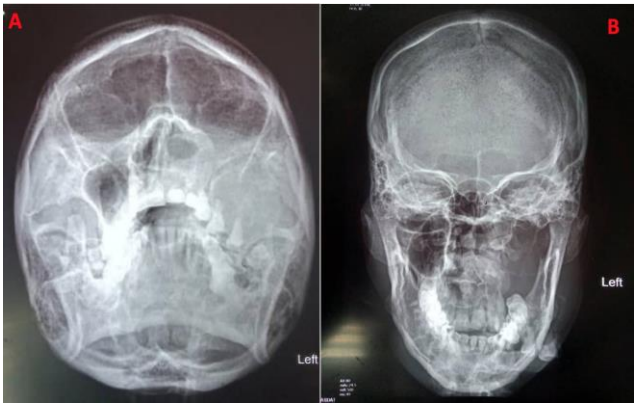

Figure (2a). Occipitomental view of the skull at 30 degrees showing radio-opacity of the left antrum with osteolysis of the antral floor, medial, medial and lateral walls and the left infra-orbital rim. There is loss of lamina dura around the 24 and 26 **(2b.)** Posterio-Anterior view of the skull showing destruction of the floor, medial and lateral walls of the antrum and ill-defined borders on the floor of the orbit

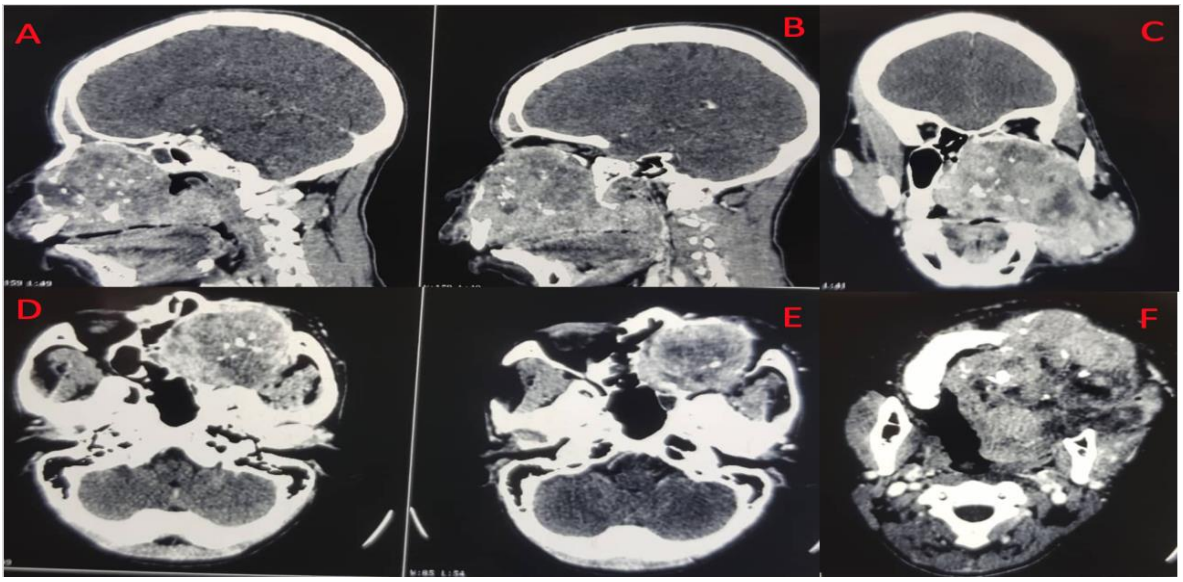

Figure 2b. Craniofacial CT Scan (soft tissue window) with views: A, B (sagittal), C (coronal), D, E and F (axial) show huge area of iso-dense mass interspersed with hyper-dense structures and extensive osteolytic destruction involving the maxilla, left maxillary antrum, nasal cavity crossing the midline, infratemporal, pterygomaxillary space and superiorly the left orbit, ethmoid and aborting the base of the skull.

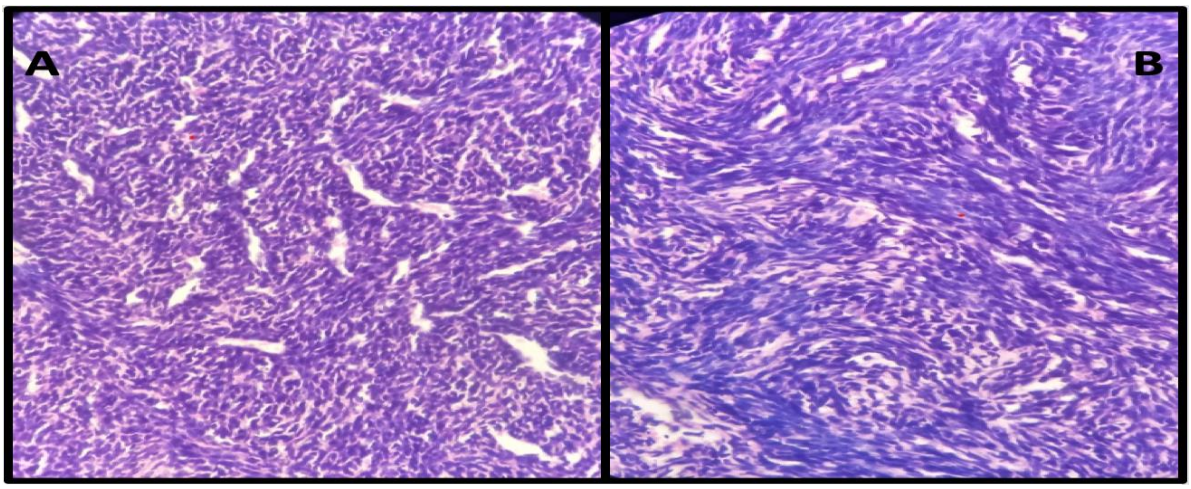

Figure (3a.) Staghorn-like vascular channels. (Hematoxylin and Eosin stain) **(3b**.) Depicting the cartwheel-like appearance towards the right and the interlacing fascicles herringbone pattern formed by the spindle cells towards the left. (Hematoxylin and Eosin, Magnification X400). There are two histologic types of fibrosarcoma: infantile and adult type fibrosarcoma.[2]

DISCUSSION

Malignant peripheral nerve sheath tumor (MPNST) is one of the differential diagnosis of this reported case.[1] On microscopic examination, MPNST shows loose connective tissue with hypo- and hypercellular areas of proliferative atypical spindle-shaped cells in streaming fascicles. Also seen are pleomorphic cells with wavy, coma-shaped and hyperchromatic nuclei.[2] Our immunohistochemistry result suggest that this case is most likely not a MPNST because it was negative for S100 and pan-cytokeratin.[3] and the neoplasm possibly had no neural origin/differentiation since it was negative for synaptophysin. A neoplasm arising from myoid or myofibroblastic origin, eg rhabdomyosarcoma

or leiomyosarcoma, was also suspected but the antibodies to myogenin and desmin were all negative so these were ruled out.

We also thought that our case had a histology that seemed somewhat biphasic, so we considered a synovial sarcoma. Synovial sarcoma is mainly seen in prearticular region close to tendon sheath and joint capsule, however there have been reported cases in the jaws.[4] Histologically, it appears as mostly monomorphic spindle cells that are moderately to highly cellular. Mild cellular pleomorphism and few atypical mitotic figures may be seen. Immunohistochemically, it stains positive for cytokeratin (AE1/AE/3)[5] and so we ruled it out, since our case was negative for this marker.

A spindle-cell melanoma was also a differential we entertained. It is a rare subtype of melanoma that mimics amelanotic lesions and could occur at any site.[6] It is composed of spindle neoplastic cells arranged in sheets and fascicles, and this makes it morphologically indistinguishable from sarcomas.[7] Our case was however negative for Melan-A and so it couldn't have been a spindle-cell melanoma.[8]

Other differentials included and eventually ruled out were hemangiopericytoma (because of the numerous irregularly shaped blood vessels reminiscent of 'stag-horn vessels'), and classic epithelioid sarcoma with areas of spindle-shaped cells but these were negative for CD34[9] and cytokeratin respectively[10,11].

Fibrosarcoma is a malignant mesenchymal neoplasm of fibroblasts that may arise from soft tissue or bone. Only 20% of cases affect bone, mainly the long bones; it rarely affects the jaw bone. Fibrosarcoma of the maxilla accounts for about 0.6.1% of all primary fibrosarcoma of bone.[12,13] There are two histologic types of fibrosarcoma: infantile and adult type fibrosarcoma[14]. Fibrosarcoma of the jaws may clinically present with swelling, pain, paresthesia and loosening of teeth.[13,15] Radiographically, it presents as an osteolytic lesion without distinct border, and cannot be distinguished from other osteolytic lesions.[16] Microscopically, it present as spindle-shaped cells with slight variability in sizes and shapes with slight nuclear pleomorphism, arranged in a herringbone, storiform or whorl pattern with increased mitosis[17].

A major challenge we had was distinguishing between fibrosarcoma and malignant fibrous histiocytoma (currently referred to as undifferentiated pleomorphic sarcoma [UPS]). UPS has been categorized into five subtypes: (a) storiform, (b) myxoid, (c) giant cell, (d) inflammatory and (e) angiomatoid. The most common is storiform UPS, typically positive for alpha smooth muscle actin (αSMA) and vimentin but negative for desmin. Fibrosarcoma on the other hand is also positive for vimentin and negative for desmin and αSMA. It is obvious to notice that the antibody needed to separate the two most likely differentials is αSMA, unfortunately however, this antibody was unavailable at the laboratory and could not be procured due to limitations with funding. The case was finally signed out as a fibrosarcoma (adult-type) rather than a storiform UPS, mostly due to morphological features. It would however had been more rewarding to also reach a final diagnosis based on immunohistochemical representation. This case highlights one of the difficulties faced in the process of diagnosis by resource-limited facilities and the possible ramifications on overall patient management. The patient presented for follow-up after 3 months (delayed in planned recall visit due to financial challenges) with a copy of the requested CT scans, this delay has further complicated the approach to management.

Conclusion: We report a case of a maxillary spindle cell neoplasm which fitted the histological and immunohistochemical profiles of a fibrosarcoma and a storiform UPS. However, the decisive marker to clearly distinguish between these two differentials was unavailable, and therefore the case was finally determined mostly by morphological features, and we can only hope that we have made the most appropriate decision for the patient and the managing team.

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