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Case Report
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Malignant Peripheral Nerve Sheath Tumor, a Sarcoma Mimicker: Importance of Cytogenetic Study

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Abstract

Isolated Peripheral Malignant Nerve Sheath Tumor (MPNST) may mimic a synovial sarcoma where a cytogenetic study is crucial to help differentiate the two entities in the context of similar clinico-histopathological background. Here we report a patient with progressive median nerve MPNST, that was initially thought of spindle cell sarcoma, however cytogenetic study excludes that possibility and patient was managed as MPNST with good outcome over 5 years follow up.

Keywords: Malignant peripheral nerve sheath tumor; Synovial sarcoma; Spindle cell sarcoma; Cytogenetic study

Introduction

Malignant Peripheral Nerve Sheath Tumors (MPNSTs) are malignant tumors which originate from peripheral nerve which had overlapping features with various sarcomas, particularly synovial sarcoma which render its diagnosis difficult and often challenging.

Synovial sarcomas are common malignant soft tissue neoplasms which can arise anywhere in the body, including peripheral nerves.

Herein, we present a case of malignant peripheral nerve sheath tumor of the median nerve which clinically, histologically and immunohistochemically mimicked synovial sarcoma, monophasic fibrous type involving the median nerve. Cytogenetic study was performed. The result did not confirm the diagnosis of synovial sarcoma, however, the possibility of a low grade MPNST was entertained. This shows the importance of cytogenetic studies in differentiating MPNST from its sarcoma mimickers.

Case Presentation

A 31-year-old previously healthy male presented with a right cubital fossa mass which was rapidly increasing in size over a 4 months period. It was very painful in the last 3 weeks prior to presentation with diffuse numbness over the right hand.

The patient gave no history of trauma, fever, weight loss or contact with an ill patient. There was no history of drug abuse.

Physical examination revealed a 10 cm × 8 cm. right cubital mass which was tense, very tender, and not pulsating nor compressible, with no radial pulsation.

Motor examination of the upper limb was 5/5, limited right elbow flexion and extension due to pain with hyposthesia mainly over right thumb and index finger.

Routine Investigation was unremarkable for infection.

CT scan of right upper limb showed Large heterogenous hyperdense cystic mass measuring 15.5 cm × 10 cm × 10 cm, likely a soft tissue mass with faint enhancement. CT angiogram showed severe compression of brachial artery and its collaterals due to mass effect.

The patient underwent emergency exploration and resection over the course of the median nerve, it was a large cystic mass encircling and intimately in close proximity to the median nerve and a brachial artery in situ graft was done by the vascular surgery team.

Histology showed a vascularized, cellular neoplasm composed of sweeping fascicles of spindly cells with scanty cytoplasm and slightly pleomorphic nuclei, less cellular areas and a

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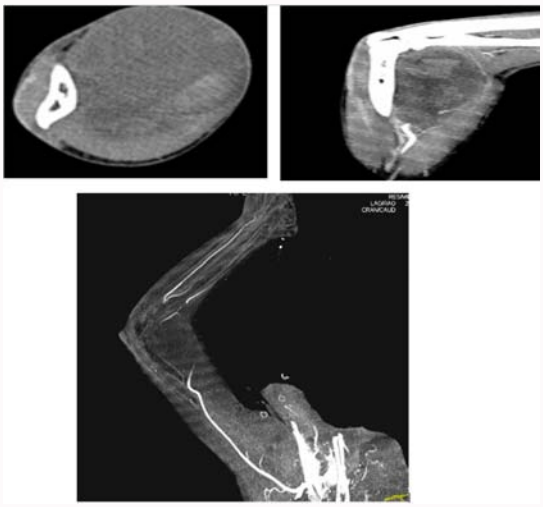

Figures 1-3: Axial, sagittal CT scan view showed a non-enhanced expansile mass occupying the right elbow region with close relation and obliteration to right brachial artery as demonstrated in Computed Tomographic Angiographic picture.

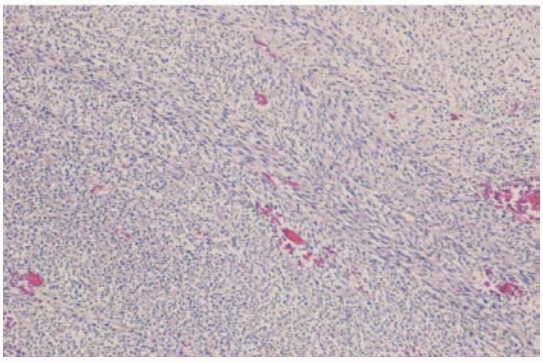

Figure 4: MPNST. Neoplastic spindly cells arranged in long fascicles. H&E x100.

"hemangiopericytoma-like" pattern were also focally observed (Figure 1,2).

Immunohistochemically, the tumor cells were diffusely and strongly positive for CD99 and CD56 (Figure 3,4). There was also focal reactivity for vimentin and S100 protein.

A diagnosis of spindle cell sarcoma was entertained (of a possible monophasic fibrous synovial sarcoma, based on operative, histologic and immunohistochemical findings).

Cytogenic study performed in the form of Fluorescence *in situ* Hybridization (FISH) analysis on paraffin-embedded tumor tissue showed no rearrangement on 18q11.2 in 200 (100%) of interphase nuclei scored. This did not confirm the diagnosis of synovial sarcoma.

Post op, the patient had symptomatic relief and regain of radial pulsation with no new deficits. He received adjunct therapy with no recurrence within a 5 years period.

Discussion

Synovial sarcomas account for 10% of soft tissue sarcomas and can involve any tissue with high prevalence in young adults [1].

Primary synovial sarcomas of nerve are very rare, with only

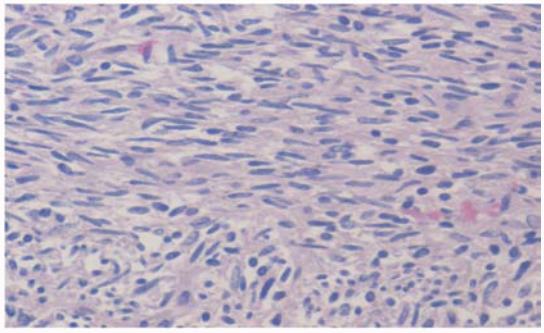

Figure 5: MPNST. High power view showing slightly pleomorphic neoplastic cells. H&E x400.

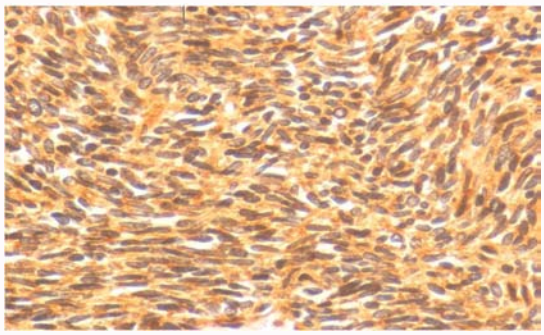

Figure 6: MPNST. Cytoplasmic immunoreactivity of neoplastic cells to CD99. IHC x400.

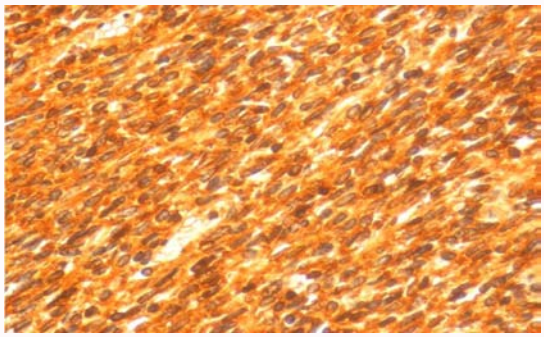

Figure 7: MPNST. Cytoplasmic immunoreactivity of neoplastic cells to CD56. IHC x400.

12 cases of intraneural synovial sarcomas of nerve reported in the literature [2,3]. The radiologic impression is that of a neurofibroma [2].

Based on analysis of the gene expression pattern, synovial sarcomas of nerve are derived mainly from epineurial connective tissue, which may originate from Schwann cells associated with the neural crest [1,4,5].

Synovial sarcomas are divided into monophasic fibrous, biphasic, purely glandular and poorly differentiated. Although biphasic morphology is most frequently reported, majority of intraneural synovial sarcomas have had monophasic fibrous histopathology [6].

The morphology of monophasic synovial sarcomas and spindle cell MPNST are very much similar. Classical features of monophasic synovial sarcoma are fascicular proliferation of monomorphic, hyperchromatic spindled cells in association with wiry collagen and

a "staghorn", hemangiopericytoma-like vascular pattern, diffuse positivity for vimentin and CD99, patchy expression of EMA, pancytokeratin and cytokeratin 7. S100 protein and CD56 are positive in a variable number of cells [7].

Translocation of t(x;18) by FISH or RT-PCR fusion transcript of SYT-SSX is the main diagnostic tool for synovial sarcomas. In contrast to its main differential diagnosis, expression of HMGA2 is a feature of MPNST [6-9].

A positive result of t(x;18) (SYT; SSX) translocation is 100% specific for synovial sarcoma, but the detection techniques used are only 86% to 96% sensitive in monophasic spindle cell synovial sarcoma. So, demonstration of the translocation confirms the diagnosis of synovial sarcoma but a negative result does not completely rule it out [10]. However, considering the overlapping clinical, radiological, operative, histologic and immunohistochemical findings, the lack of rearrangement on 18q11.2 as revealed by FISH and the location of the tumor in relation to a large nerve favored the diagnosis of MPNST over synovial sarcoma.

Conclusion

A rapidly growing spindle cell tumor of a peripheral nerve should raise the suspicion of a highly malignant tumor, with a differential diagnosis of peripheral malignant nerve sheath tumor *vs.* sarcoma. Monophasic fibrous synovial sarcoma should always be considered. Achieving a correct diagnosis in the context of cytogenetic study is of crucial importance, considering the overlapping clinical, radiological, operative, histologic and immunohistochemical findings between MPNS and synovial sarcoma.

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