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Case Report

Pleomorphic rhabdomyosarcoma of lumbar region: A case report

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ABSTRACT

Rhabdomyosarcoma is a malignant mesenchymal soft tissue tumour with tendency towards skeletal muscle differentiation. It is commonly seen in children and adolescents but is exceedingly rare in adults. The most common site in adults is the extremities. Less common locations include pelvis and thorax. The tumour is diagnosed by the morphologic presence of scattered pleomorphic rhabdomyoblasts and immunohistochemistry (with evidence of at least one skeletal muscle-specific marker). The outcome of tumour in adults is very poor, especially when compared to outcomes in children in whom significant improvements with treatment have been achieved. We present a case of Pleomorphic Rhabdomyosarcoma at the lumbar region which was clinically thought as a dermoid cyst. This case is reported because of rare location. The diagnosis was made on histopathology and immunohistochemistry. The tumour was diffusely positive for vimentin and desmin and focally positive for myogenin & MyoD1.

1. Introduction

Rhabdomyosarcoma is a primitive malignant mesenchymal tumour with tendency for striated muscle tissue differentiation.[1] It is the most common type of childhood soft tissue sarcoma, comprising 5 to 10% of all solid tumours in childhood. However, it rarely occurs in adults; soft tissue sarcomas account for less than 1% of all cancers in adults.[2,3] Rhabdomyosarcoma is recognized as four subtypes: alveolar, embryonal, pleomorphic and spindle cell/sclerosing.[4] Alveolar and embryonal subtypes of rhabdomyosarcoma are the most common soft tissue sarcoma of childhood and adolescence, usually appearing before 20 years of age, while pleomorphic rhabdomyosarcoma occurs in adults. Pleomorphic rhabdomyosarcoma is composed of large, multinucleated, bizarre eosinophilic tumour cells that can overlap histologically with other pleomorphic sarcomas. Immunohistochemistry (myogenin or MyoD1) is usually necessary to confirm rhabdomyoblastic differentiation.[5] We report a case 79-year-old male with a lump in the lumbar region which was clinically suspected to be a dermoid cyst.

2. Case Report

A 79- year-old male, presented with a lump in the back since 2 years. On examination, a 6x5cm lump was seen in the right lumbar region. The lump was clinically suspected to be a dermoid cyst and thus no radiological investigation was done. The lump was later excised and sent for histopathological examination. On gross examination, external surface appeared brownish and nodular. Cut surface showed a greyish white and fleshy growth measuring 5x5x2.5cm with a peripheral rim of brown normal looking tissue (Figure 1 A).

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Sections were stained with H&E and studied. Microscopy showed tumour composed of cells having round to oval nucleus with prominent nucleoli and moderate eosinophilic to clear cytoplasm. There was moderate to marked nuclear pleomorphism and increase in mitosis. Wide areas of necrosis and haemorrhage were noted constituting about 25-30% of tissue (Figure 1 B, C, D). Large rhabdomyoblasts with irregular hyperchromatic nucleus and deeply eosinophilic cytoplasm were seen.

Immunohistochemistry showed strong and diffuse positivity for Desmin, positive for Vimentin and focal positivity for Myogenin and MyoD1. Ki- 67 showed high index positivity (Figure 2). Other malignant sarcomas were also ruled out by IHC panel [Pan CK, SMA, S100, H-caldesmon, HMB-45, Sox10, CD34 and ERG and were negative]. Thus, sarcomas of epithelial, smooth muscle, melanocytic, fibrous & vascular origins were excluded (Figure 3). Histopathology along with IHC led to the diagnosis of Pleomorphic Rhabdomyosarcoma. Patient was discharged the next day and his condition was satisfactory.

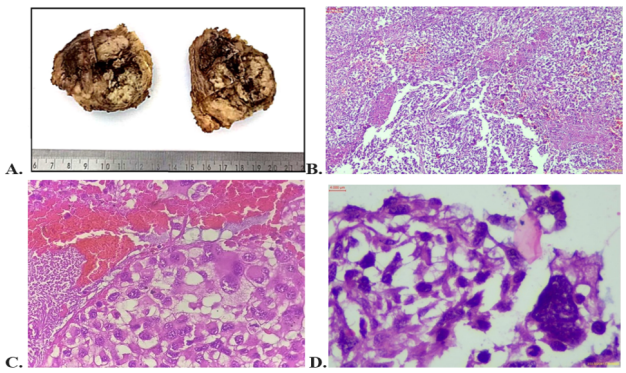

Fig. 1: A): Gross image, **B**): Haematoxylin and eosin stain (HE) 4X; **C**): H&E stain 40×, **D**): H&E stain 100X

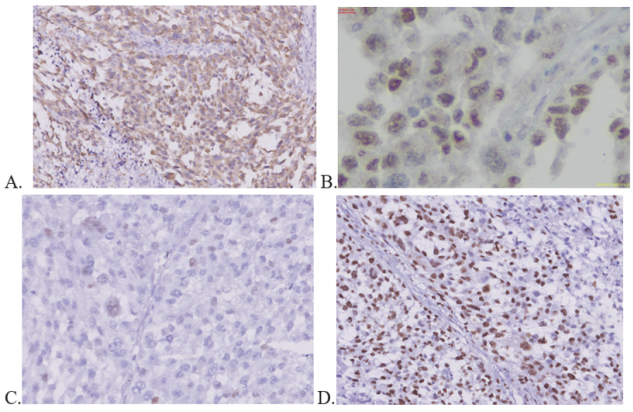

Fig. 2: Immunohistochemistry **A**): Desmin 100x, **B**): MyoD1 100x, **C**): Myogenin 100x, **D**): Ki-67 100x

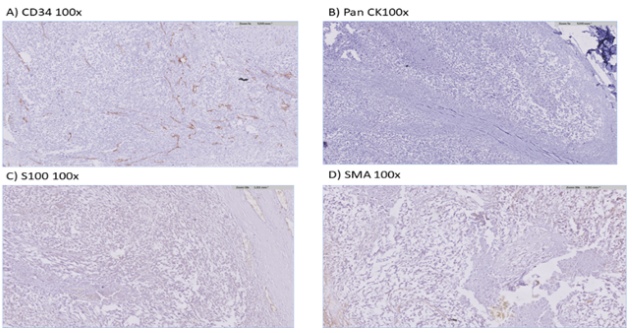

Fig. 3: Immunohistochemistry **A**): CD34 100x, **B**): Pan CK 100x, **C**): S100 100x, **D**): SMA 100x

3. Discussion

Pleomorphic Rhabdomyosarcoma was first introduced into literature by Stout in 1947 as classical rhabdomyosarcoma.[1] For the next three decades diagnosis was based on morphology alone and probably included other sarcomas. In 1958, Horn and Enterline outlined four subtypes of rhabdomyosarcoma. The classical ones were called pleomorphic rhabdomyosarcoma.

Pleomorphic Rhabdomyosarcoma is a high-grade pleomorphic sarcoma of skeletal muscle differentiation. This category of tumour is highly aggressive and exceeding rare in adults. It is usually associated with poor prognosis. It usually presents in the 6^{th} to 7^{th} decade, with a male predominance of 1.8:1. Usual site of presentation is deep soft tissues of the extremities. Less common locations include pelvis, abdomen, thorax, head and neck.[6] Rare cases of the tumor have been reported in liver[3] and chest wall infiltrating intercostal muscles.[7] It is usually a rapidly growing painful mass, but mass since 3years also have been reported.[7] Grossly they are usually large, well-marginated tumours, with a pseudo capsule and a whitish or fleshy cut surface, often with necrosis. The tumours are composed of sheets of large, atypical, and frequently multinucleated polygonal, spindled, or rhabdoid cells with eosinophilic cytoplasm.

Immunohistochemical antibodies are applied to these tumours to detect skeletal muscle differentiation.[1] Myogenin is the most sensitive and specific marker of rhabdomyosarcoma. The myogenin gene codes for a phosphoprotein that induces skeletal muscle differentiation in mesenchymal cells. The protein can be demonstrated in the nuclei of the tumour cells in all cases of rhabdomyosarcoma.[5] MyoD1 is a related nuclear protein with similar degree of specificity. Other markers include desmin, sarcomeric actin, myosin, myoglobin, titin, and Z-protein. Some are less useful and not frequently used in clinical practice.[5] Pleomorphic Rhabdomyosarcomas typically display strong desmin expression and often limited

expression of MyoD1 and myogenin.

Pleomorphic Rhabdomyosarcoma typically resembles an undifferentiated pleomorphic sarcoma except for the presence of scattered pleomorphic rhabdomyoblasts and myogenin immunoreactivity.[5] Differential diagnosis includes liposarcoma (presence of lipoblasts, S100+), fibrosarcoma (spindle cells in herring bone pattern), angiosarcoma (ERG+, CD34+, CK+/-), myxofibrosarcoma (spindle cells in a myxoid background, Desmin -), malignant peripheral nerve sheath tumor (spindle cells with alternating hyper and hypocellular areas, can have rhabdosarcomatous component, S100+, SOX10+), spindle cell squamous cell carcinoma (CK+, EMA+) and desmoplastic melanoma (S100+, SOX10+).[8] Based on morphology and immunohistochemistry these were ruled out.

4. Conclusion

We presented a case of a 79- year old male with lump in the Right lumbar region since 2 years. The case is presented here because pleomorphic rhabdomyosarcoma of lumbar region is very rare. IHC marker myogenin and MYOD1 is focally positive which indicates skeletal muscle differentiation. IHC also helps to differentiate from melanoma and other undifferentiated pleomorphic sarcomas. Patient was discharged the next day and his condition is satisfactory.

5. Source of Funding

None.

6. Conflict of Interest

None.

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