A Rare Case of Hilar Lymph Node Schwannoma

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Abstract Schwannoma arising in a lymph node is an extremely rare primary mesenchymal tumor of the lymph node. Although intranodal schwannoma is benign, this lesion in a lymph node may be initially alarming and it may suggest a diagnosis of intranodal metastasis from a spindle cell tumor. Microscopic examination of tumor shows Antoni A areas, which are moderate to highly cellular areas of elongated cells arranged in fascicles, and Antoni B areas, which are hypocellular areas with myxoid and microcystic change. Tumor cell morphology is similar in both areas with elongated shape and regular oval nuclei. Immunohistochemistry report shows strong and diffuse positivity for S-100 which is consistent with Schwann cell origin. We report a rare case of intranodal schwannoma arising in the hilar lymph node with its clinical, light microscopic, and immunohistochemical findings

Keywords Schwannoma · Lymph node · Myofibroblastoma

Case Details

A 42-year-old female patient presented with low-grade fever, dry cough, and dyspnea of 18 day's duration. X-ray chest showed prominence of right hilum and a suspected mass lesion. Subsequently, a CT scan chest was performed, which showed a 39 mm×36 mm space-occupying lesion (SOL) with well-defined margins in right paravertebral region. No erosion of underlying ribs/vertebra was seen. Mild enhancement with contrast was seen and a strong possibility of neurofibroma was considered. MRI of the dorsal spine showed a right paravertebral mass lesion, measuring 46 mm×

36 mm×24 mm, involving the D7 vertebral region. The lesion was isointense to muscle on T1-weighted images and heterogeneously hyperintense on T2-weighted images (Fig. 1). No pressure effects were noted on adjoining structures (vertebrae and ribs). Ipsilateral adjacent pleura was thickened. Biochemical parameters were within normal limits: alkaline phosphatase 44 IU/l, serum calcium 9.5 mg/dl, and serum phosphorus 2.8 mg/dl. A provisional diagnosis of the benign soft tissue lesion of neurogenic origin was given and the patient underwent excision of the mass lesion.

Grossly the excised mass was a single whitish nodule, measuring 4.2 cm×3.5 cm×2.4 cm, which on the cut section had a homogenous fleshy appearance. Sections from the tumor submitted showed a well-encapsulated lymph node exhibiting effacement of architecture by a spindle cell lesion showing cellular areas with intersecting and criss-cross fascicles admixed with variable amount of collagen. Compressed normal lymphoid tissue was seen at the periphery. Areas showing nuclear palisading and thickened hyalinized blood vessels were present. Scattered lymphoid follicles with prominent germinal centers and organoid structures resembling Pacini's corpuscles were noted (Fig. 2). Immunohistochemistry showed vimentin and S-100 to be strongly positive. Leukocyte common antigen (LCA) (CD45) was strongly expressed in lymphoid component. Desmin, smooth muscle actin (SMA), human melanoma black (HMB45), cytokeratin (CK), and epithelial membrane antigen (EMA) were all negative. A diagnosis of lymph node Schwannoma was given correlating the histopathological and immunohistochemical findings.

Discussion

Intranodal schwannoma is an extremely rare benign lesion [1–3]. A very few genuine cases of benign schwannoma arising in a lymph node are known. Most

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Fig. 1 **a** X-ray chest PA view shows hilar prominence and suspected mass lesion. **b** CT scan chest shows a 39 mm×36 mm space-occupying lesion (SOL) with well-defined margins in right paravertebral region. **c** MRI dorsal spine shows a right paravertebral mass lesion measuring 46 mm×36 mm×24 mm involving the D7 vertebral region

of lesions previously diagnosed have nowadays been recognized as intranodal myofibroblastomas [4]. In 1989, Weiss et al. reclassified their collection of 22

cases including many lesions that were diagnosed as intranodal schwannoma as palisaded myofibroblastoma [5]. As in other reported cases [1–3, 6] of intranodal schwannoma, our case also had a central tumor with typical features of a soft tissue schwannoma rimmed by compressed normal lymphoid tissue. A peculiar finding in our case was the presence of organoid structures resembling Pacini's corpuscles. Lesions considered in the differential diagnosis of intranodal schwannoma are metastatic spindle cell tumors including sarcomas, spindle cell carcinoma, and melanoma. Among the benign spindle cell intranodal lesions, most important differential diagnosis is palisaded intranodal myofibroblastoma. Features which differentiate intranodal schwannoma from palisaded myofibroblastomas are extensive foci of hemorrhage and stellate areas of collagen deposition, the so-called amianthoid fibers seen in latter. These lesions show smooth muscle actin positivity on immunohistochemistry but not S-100 expression which is strongly positive in schwannomas [5, 7]. Our case was strongly positive for S-100 and negative for smooth muscle actin, thereby ruling out intranodal myofibroblastoma. CK and EMA were negative ruling out metastatic spindle cell carcinoma, and HMB45 negativity ruled out a metastatic spindle cell melanoma.

Among soft tissue sarcomas metastatic to lymph nodes, synovial sarcoma, rhabdomyosarcoma, and angiosarcoma are relatively more frequent and malignant schwannomas do so very infrequently. Tumor cells exhibit cytological atypia and are mitotically active and are different from bland-looking spindle cells of a schwannoma [8]. Monophasic synovial sarcoma is hypercellular and remarkably monomorphic. Expression for keratins and EMA can be seen at least focally.

In follicular and interdigitating dendritic cell sarcomas, neoplastic cells are generally plumper than schwannoma cells and usually form nests and whorls [9]. Unlike schwannoma, they express LCA, CD35, and CD68. Primary Kaposi sarcoma of the lymph node may show relatively bland-looking spindle cells, but the presence of slits containing erythrocytes, hemosiderin pigment, and immunohistochemistry (IHC) profile (CD34 positive) enables ruling out schwannoma [10].

In conclusion, we have reported an extremely rare case of schwannoma arising in a lymph node. Because of its rarity, it is a diagnostic challenge and other primary and metastatic spindle cell lesions must be excluded before giving this diagnosis.

References

1. Griffiths AP, Ironside JW, Gray C (1991) True neurilemmoma arising in a lymph node in infancy. *Histopathology* 18(2):180–183

Fig. 2 **a** Low power view of the mass lesion shows encapsulated lymph nodes with peripheral rim of compressed lymphoid tissue and central schwannoma with Antoni A area. **b** Organoid structures resembling Pacini's corpuscles. **c** Schwannoma: Antoni B area and lymphoid follicles. **d** Immunohistochemistry: strong S-100 positivity of tumor cells

- Piana S, Gelli MC, Cavazza A, Serra L, Gardini G (2002) Ancient schwannoma arising in a lymph node: report of a case and review of literature. *Pathol Res Pract* 198(1):51–54
- Reinus C, Shutin O, Reissman P (2004) Intranodal schwannoma presenting as an adrenal mass. *Histopathology* 45(1):95–96
- Fletcher CD (2000) Primary neuroectodermal tumours. In: *Diagnostic histopathology of tumours*. Churchill Livingstone, p 1686
- Weiss SW, Gnepp DR, Bratthauer GL (1989) Palisaded myofibroblastoma. A benign mesenchymal tumour of lymph node. *Am J Surg Pathol* 13:341–346
- Jung JH, Jinyoung Y, Seok JK, Kyo YL (2009) Schwannoma arising in a lymph node. *Korean J Pathol* 43:217–213
- Suster S, Rosai J (1989) Intranodal haemorrhagic spindle-cell tumor with “amianthoid” fibres. Report of six cases of a distinctive mesenchymal neoplasm of the inguinal region that simulates Kaposi’s sarcoma. *Am J Surg Pathol* 13:347–357
- Nasrin S, Shahriar ZS (2008) Schwannoma in a perigastric lymph node: a rare case report. *Iran J Pathol* 3(1):43–46
- Hollowood K, Pease C, Mackay AM, Fletcher CD (1991) Sarcomatoid tumors of lymph nodes showing follicular dendritic cell differentiation. *J Pathol* 163(3):205–206
- Chan JKC (1997) Vascular tumors with a prominent spindle cell component. *Curr Diagnost Pathol* 4(2):76–90