CASE REPORT

Reticular schwannoma mimicking myxoid sarcoma

Jai Kumar Chaurasia,¹ Nishat Afroz,¹ Biswajit Sahoo,² Mohammed Naim¹

¹Department of Pathology, Jawaharlal Nehru Medical College (JNMC), AMU, Aligarh, Uttar Pradesh, India
²Department of Radiodiagnosis, Jawaharlal Nehru Medical College, Aligarh Muslim University (AMU), Aligarh, Uttar Pradesh, India

Correspondence to

Dr Jai Kumar Chaurasia,
 dr.jaichaurasia@gmail.com,
 dr.jaichaurasia@yahoo.com

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SUMMARY

Reticular/microcystic schwannoma is a recently described rare distinctive variant of schwannoma with a predilection for gastrointestinal tract. Its occurrence in soft tissue is extremely rare. We report a case of reticular/microcystic variant of schwannoma in a 55-year-old Indian woman who presented with a painless slow-growing swelling in the right forearm for the past 6 months. MRI findings suggested myxoid sarcoma. However, histological and immunohistochemical findings in the excised specimen were consistent with reticular/microcystic variant of schwannoma. This case report emphasises that the diagnosis of reticular/microcystic schwannoma should always be considered in cases where myxoid sarcomas are suspected as it can mimic malignant myxoid sarcomas, clinically and radiologically, thereby avoiding aggressive intervention and overtreatment.

BACKGROUND

Schwannomas (neurilemmomas) are common benign peripheral nerve sheath tumours composed of a relatively uniform population of cells showing schwannian differentiation. Many variants including ancient, plexiform, cellular, melanotic, gastric and epithelioid schwannomas have been described with a wide range of histomorphologies. The reticular/microcystic variant of schwannoma is a recently described rare distinctive morphological entity with a predilection for visceral locations in the gastrointestinal tract.^{1–3} We report a rare case of reticular/microcystic variant of schwannoma mimicking a soft tissue myxoid sarcoma.

CASE PRESENTATION

A 55-year-old Indian woman presented with the symptoms of painless slow-growing swelling in the right forearm for the past 6 months. There was no significant family history or hereditary predisposition. No history suggestive of systemic infection, any autoimmune disease or trauma at the site of swelling was present. Clinical examination revealed a well-defined, soft-to-firm, non-tender swelling, measuring 5×4 cm in size, involving the distal one-third and wrist of the volar aspect of the right forearm. The swelling was immobile along the long axis of the forearm. Overlying skin was normal without any signs of inflammation. No axillary or cervical lymphadenopathy was appreciated. Chest X-ray and CT scan of the abdomen were found to be unremarkable. Fine-needle aspiration cytology (FNAC) of the swelling yielded only myxoid material. An MRI revealed a well-defined, altered signal intensity lesion, measuring 5×4.5×2.5 cm in the subcutaneous plane on the posteromedial aspect of the lower arm, abutting and displacing the triceps

laterally with indistinct intervening fat planes. The swelling was homogeneously hyperintense on T2-weighted MRI sequences (figure 1A). Postcontrast T1-weighted MRI showed heterogeneous enhancement of the lesion with hyperintense central and hypointense peripheral part (figure 1B). The MRI findings were suggestive of a myxomatous tumour, favouring a low-grade fibromyxoid sarcoma (LGFS) or an extraskeletal myxoid chondrosarcoma (EMCS). In view of the MRI findings suggesting malignancy, the equivocal clinical picture and non-contributory FNAC, a diagnosis of low-grade myxoid sarcoma was considered. The swelling was surgically excised and submitted for histopathological examination. Gross examination of the specimen revealed an encapsulated, round-to-oval, soft-to-firm tissue mass, measuring 6×5×3 cm with shiny white outer surface (figure 1C) and yellow-white cut surface (figure 1D). Whole tissue was processed and paraffin blocks were prepared.

H&E-stained sections showed partially encapsulated lesion with a characteristic reticular microcystic growth pattern with intersecting strands of spindle cells arranged around degenerating myxoid stroma (figures 2 and 3). The tumour cell morphology varied from round, oval-to-spindle shape with slender wavy nuclei (figure 2). Degeneration in the form of extensive myxoid change was also seen with sclerosis and thick-walled blood vessels in peripheral areas (figures 4 and 7). Prominent nuclear palisading (Verocay bodies) was appreciated focally (figure 5). No areas of necrosis were present. Nuclear atypia or mitosis was not appreciable. Immunohistochemical examination demonstrated a strong diffuse positivity of spindle cells to S-100 (figure 6) and vimentin (figure 7) and non-reactivity to epithelial membrane antigen (EMA), smooth muscle actin (SMA), desmin, P-16, CD-117, CD-34, CD-45, cytokeratin and synaptophysin. Based on histological and immunohistochemical findings, a final diagnosis of benign nerve sheath tumour consistent with a reticular/microcystic variant of schwannoma with extensive myxoid degeneration was made.

INVESTIGATIONS

Routine haematological, urine examination and biochemical tests of the patient were normal.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes LGFS, EMCS, reticular perineurioma, myoepithelial tumour, ganglioneuroma with abundant myxoid stroma, myxoid variant of dermatofibrosarcoma protuberans (DFSP), myxoid malignant peripheral nerve sheath tumour (MPNST) and synovial sarcoma with extensive myxoid changes. The findings, in

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Figure 1 (A) T2-weighted MRI showing homogeneously hyperintense lesion (arrow), (B) postcontrast T1-weighted MRI showing heterogeneous enhancement of the lesion with hyperintense central and hypointense peripheral part (arrow), (C, D) gross specimen showing round-to-oval tumour, with shiny white outer surface and yellow-white cut surface (arrow) respectively.

the present case, were diagnostic of reticular schwannoma, distinguishable from these tumours by its distinctive histological and immunohistochemical features.

OUTCOME AND FOLLOW-UP

The postoperative recovery of the patient was uneventful. No recurrence of the tumour was seen during the follow-up period of past 2 months.

DISCUSSION

Reticular/microcystic schwannoma is a rare variant of schwannoma and its occurrence in soft tissue, as was seen in the present case, is extremely rare. So far, only Liegl *et al*¹ have reported two cases of reticular schwannoma in subcutaneous location in the year 2008.

Reticular schwannoma can mimic an LGFS as it may share overlapping clinical and radiological features. An LGFS is an uncommon slow-growing soft tissue tumour, clinically presents as painless, well-circumscribed soft tissue mass and can occur in dermis or subcutaneous tissue.⁴ Radiologically, it shows typical findings of myxoid neoplasm, that is, homogeneously hyperintense lesion on T2-weighted MRI and heterogeneous postcontrast enhancement with hyperintense central part, due to areas of myxoid degeneration in the tumour and hypointense peripheral part, similar to the present case of reticular schwannoma, causing diagnostic difficulty.⁵ Presently, although FNAC was not diagnostic and yielded only myxoid material, it raised the suspicion of a myxoid neoplasm. However, histological and immunohistological examinations distinguished these two entirely

Figure 2 Tumour section showing characteristic reticular/microcystic growth pattern with intersecting strands of spindle cells (H&E ×50).

Figure 3 Tumour encapsulation (arrow) with underlying myxoid degeneration (H&E ×50).

Figure 4 Tumour section with myxoid degeneration (arrow) and thick-walled blood vessels (double arrow) in peripheral area (H&E $\times 50$).

different entities. Histologically, LGFS exhibits identifiable alternation of myxoid and fibrous or collagenous zones with bland spindle cells in whorled pattern. The myxoid zones have prominent curvilinear and branching arcade blood vessels, and, occasionally, giant collagen rosettes. Immunohistochemically, although LGFS show vimentin positivity, a strong immunoreactivity for S-100 in spindle cells, as seen in reticular schwannoma, is absent. Moreover, focal immunoreactivity for muscle markers such as SMA can be present in LGFS, suggesting myofibroblastic differentiation of its tumour cells. P-16 non-reactivity, in the present case, excluded the possibility of low-grade sarcoma or any sarcomatous transformation in the lesion.

EMCS is also a uncommon slow-growing soft tissue neoplasm and is considered as a unique entity of uncertain differentiation, distinct from skeletal myxoid chondrosarcoma, as it shows paucity of convincing evidence of cartilaginous differentiation.⁶ Clinically and radiologically, it can also simulate a reticular schwannoma as it usually presents as soft-to-firm, well-circumscribed, round-to-oval mass and show features of myxoid neoplasm on MRI. However, histologically, EMCS has a characteristic multinodular pattern with round or elongated cells of uniform shape with small hyperchromatic nuclei and a narrow rim of deeply eosinophilic cytoplasm, reminiscent of

Figure 5 'Verocay bodies' with palisaded tumour cells (H&E $\times 125$).

Figure 6 Strong S-100 positivity of tumour cells ($\times 50$).

chondroblasts, separated by variable amounts of mucoid material. Prominent nuclear palisading (Verocay bodies) typically of a schwannoma is absent. Strong immunoreactivity of spindle cells for S-100 in the present case of reticular schwannoma distinguished it from EMCS that shows only weak or absent S-100 immunoreactivity.

Soft tissue reticular perineurioma is a rare extraneural perineurioma which occurs as painless, solitary, well-circumscribed subcutaneous swelling or nodule. Radiologically, it also shows characteristic features of a myxoid neoplasm.⁷ Histologically, it has spindle cells with bipolar cytoplasmic processes, forming a reticular or net-like growth pattern within myxoid stroma. However, immunohistochemically, it is distinguishable from reticular schwannoma by S-100 non-reactivity and diffuse reactivity to EMA marker.⁸

Soft tissue myoepithelial tumour is also well-circumscribed tumour and histologically shows reticular architecture with nests, cords and trabeculae of epithelioid and spindle cells within a chondromyxoid to hyalinised stroma.⁹ Immunohistochemical positivity of its tumour cells to cytokeratin, EMA and SMA distinguishes myoepithelial tumour from reticular schwannoma.

Ganglioneuroma with abundant myxoid stroma also shows radiological features of homogeneous marked hyperintense

Figure 7 Vimentin positivity of tumour cells with thick-walled blood vessels (arrow; $\times 50$).

Learning points

- ▶ The reticular/microcystic schwannoma is a recently described rare distinctive novel variant of schwannoma with a predilection for gastrointestinal tract. Its location in soft tissue is extremely rare.
- ▶ The diagnosis of reticular/microcystic variant of schwannoma should always be considered in cases where myxoid sarcomas are suspected, as it can mimic malignant myxoid sarcomas, clinically and radiologically, thereby, avoiding aggressive intervention and overtreatment.
- ▶ This case report further affirms the importance of histological and immunohistochemical examination for correctly diagnosing this rare reticular variant of schwannoma.

lesion on T2-weighted MRI, similar to a reticular schwannoma.¹⁰ However, it was excluded in the present case, as no ganglion cells were seen histologically.

Myxoid variant of DFSP may histologically resemble reticular schwannoma, but the cells of DFSP are arranged in a storiform pattern and consistently stain for CD34 in contrast to the present case.

MPNST is distinguishable from reticular schwannoma as it histologically demonstrates substantial nuclear atypia, significant mitotic figures, perivascular hypercellularity and tumour necrosis. The reticular architecture is generally not seen in MPNST. Moreover, MPNST shows only focal weak S-100 immunopositivity. Similarly, synovial sarcoma with extensive myxoid changes

can be differentiated histologically by the presence of fascicular spindle cell areas, nuclear atypia, high mitotic index and positive EMA and cytokeratin immunorexpression.

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Competing interests None.

Patient consent Obtained.

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