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Case Report

A microcystic/reticular schwannoma in an unusual site: description of a retroperitoneal location and review of the literature

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Summary

Microcystic/reticular (MRV) schwannoma has been described since 2008, but remains a rarely encountered entity. MRV has a predilection for visceral locations and has variable histologic appearances. Given its rarity and anatomic variability, this entity could raise differential diagnostic issues with other tumours and malignancies.

We describe the case of a 69-year-old male followed at IRCCS Ospedale Policlinico San Martino of Genoa for his previous history of non-Hodgkin lymphoma. A para-aortic mass was discovered during follow-up, which -due to its stability, also after chemotherapy- had been hypothesized to be a non-lymphomatous lesion; given the dimensions and the site, the mass was removed. Histological evaluation showed a nodule limited by a slight fibrous capsule and characterized by a proliferation of medium-sized fusiform cells, with elongated nuclei and scarce eosinophilic cytoplasm. Given the lack of malignant signs and the strong expression of protein S-100, a diagnosis of mesenchymal neoplasia with expression of neural markers compatible with reticular schwannoma was made. The neoplasm has not recurred since its removal.

The case we present is, at our best knowledge, the first described in the retroperitoneum, a site where the exclusion of other mesenchymal malignancies is mandatory. The rarity and variability of presentations could create problems of differential diagnosis both with mucinous-producing carcinomas or with other soft tissue tumours, with myxoid or reticular structure. The description of this case could help raise information on this rare neoplasm and help distinguish it from other malignancies, especially in unusual sites.

Key words: reticular schwannoma, pathology, soft tissue tumor, rare neoplasms

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Introduction

Schwannomas are benign tumors of the nerve sheath, typically solitary and with wide anatomic distribution. Morphologic variants of schwannomas include ancient, plexiform, cellular, epithelioid and melanotic [1-3].
A more recently identified variant is the microcystic/reticular (MRV) schwannoma, described for the first time in 2008, and subsequently described in some case reports, although remaining a rare encountered entity [1-12].
In contrast to other variants of schwannomas, MRV has a predilection for visceral locations and it is histologically characterized by a striking microcystic and reticular growth pattern, composed of anastomosing and intersecting spindle cells admixed with vacuolated cells within a collagenous to myxoid stroma [1-2].

Due to its rarity and anatomic variability, this entity could raise differential diagnostic issues with other tumors and malignancies.
We here describe a case of microcystic/reticular schwannoma, developed in the retroperitoneum of a 69-year-old male, in order to help collect other information on this tumor.

Case presentation

A 69-year-old male was followed at Ospedale Policlinico San Martino of Genoa because of his previous history of non-Hodgkin lymphoma. The evaluation of CT scans during and following chemotherapy showed a dishomogeneous para-aortic mass initially of 5 centimeters, which – due to its stability for five years, also after chemotherapy – had been hypothesized to be a non-lymphomatous lesion. Radiologically the mass was suspected to be a paraganglioma or a neuroendocrine tumor.
The patient underwent the surgical excision of the mass, which macroscopically resulted to be a nodular formation of 10 cm of maximum diameter, partly covered with adipose tissue. The cut surface was dishomogeneous, yellowish, with translucid and hemorrhagic areas.
Hematoxylin-eosin sections were obtained from numerous paraffin-embedded inclusion blocks and subsequent immunohistochemical reactions for S100, EMA, CD34, CD31, MDM-2, Melan-A, HMB45, Smooth Muscle Actin, Desmin, Calponin, Synaptofisin, Chromogranin, CD117, HHV-8, Ki-67 and Cytokeratins AE1-AE3 and CAM5.2 were performed (Ventana Benchmark).
Histological evaluation showed a circumscribed mass, peripherically limited by a slight fibrous capsule (Fig. 1) and characterized by a proliferation of medium-sized fusiform cells, with elongated nuclei and scarce eosinophilic cytoplasm (Fig. 2).
Neoplastic elements had various pattern of growth: some areas showed a tight proliferation, while the

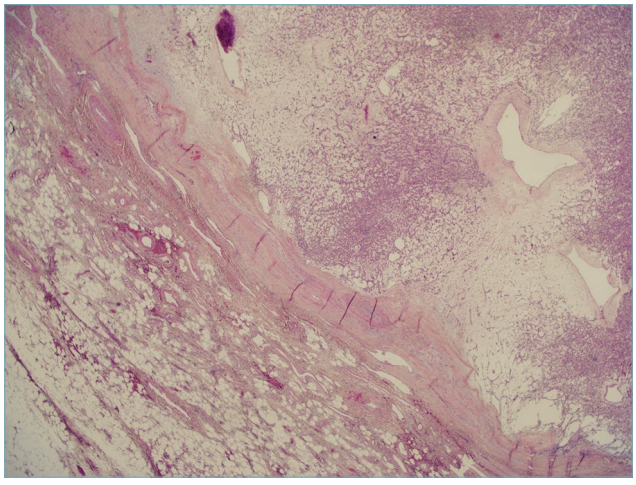

Figure 1. Hematoxylin-Eosin 20x: a slight fibrous capsule divides the neoplastic mass (on the right) from retroperitoneal fibro-adipose tissue.

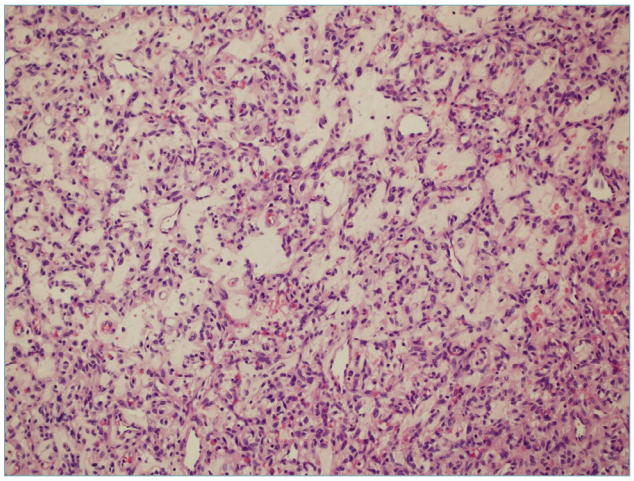

Figure 2. Hematoxylin-Eosin 200x: the neoplasia was composed of elongated bland cells, in a microcystic/reticular structure; spaces between neoplastic cells were filled with focally myxoid and hemorrhagic content. Numerous intermingled capillaries can be seen.

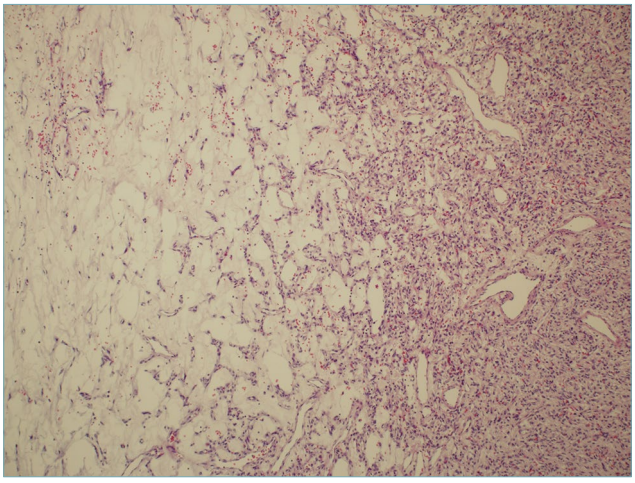

Figure 3. Hematoxylin-Eosin 100x: medium enlargement of different parts of the mass, with a tighly packed area (on the right) and a microcystic/reticular structure on the left.

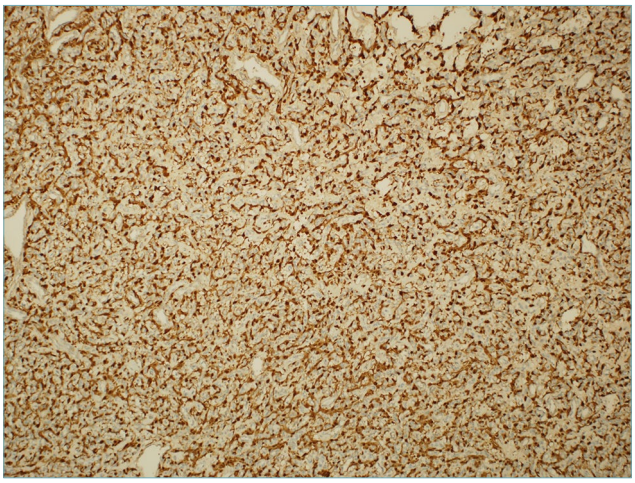

Figure 4. Immunohistochemical stain for S100: a strong and diffuse positivity from neoplastic cells was highlighted, confirming the nervous nature of the cells.

majority of the lesion had partly a microcystic or a reticular structure, depending on the width of space between the anastomotic strands of fusiform cells (Fig. 3). Spaces between neoplastic cells were filled with focally myxoid and haemorragic content.
Numerous vascular structures were mixed with neoplastic cells. No necrosis or mitotic figures were found. No aggregations of lymphocytes or other inflammatory cells were identified.
Immunohistochemical evaluation highlighted a strong and diffuse expression for protein S-100 in fusiform elements (Fig. 4), with a weak reaction for EMA in the capsule. Vascular markers CD31 and CD34 were expressed in intranodular vessels. Cytokeratins (AE1-AE3 and CAM5.2) and muscular, neuroendocrine and melanocitic stainings (Smooth Muscle Actin, Desmin, Calponin, Synaptofisin, Chromogranin, CD117, MDM-2, Melan-A, HMB45 and HHV-8) were all negative.
Given the lack of malignant signs and the strong expression of protein S-100, a diagnosis of mesenchymal neoplasia with expression of neural markers compatible with reticular schwannoma was made.

Discussion

Schwannomas are benign tumors of the nerve sheath, typically solitary and with wide anatomic distribution, usually arising in the fourth or fifth decade of life, in the subcutaneous tissue of distal extremities, or in the head and neck region of adult patients, with no gender predilection [4].

Among the numerous possible variants of this tumor, in 2008 Liegl et al. first described the microcystic/reticular schwannoma [1,2]. This variant has a predilection for visceral locations, especially gastrointestinal tract, and a slight predilection for women, with a wide age range distribution [4,5].
Histologically it is usually characterized by a striking microcystis and reticular growth pattern, formed by anastomosing and intersecting spindle cells admixed with vacuolated cells within a collagenous to myxoid stroma; scattered Verocay bodies are evident. Tumor cells have spindle to vacuolated shape and ovoid nuclei, inconspicuous nucleoli, and eosinophilic cytoplasm. Foci of classical schwannoma patterns (Antoni A and Antoni B areas) are often present. Scattered inflammatory cells could be present, but lymphoid aggregates are rare. Nuclear atypia, necrosis, and mitotic activity are rare or absent [1,2].
Since 2008, a total of 36 cases have been described [1-12], all confirming the benign nature of the lesion and showing the variability of possible presentations. The majority of cases arose in the in gastrointestinal tract, both upper and lower segments, but subcutaneous fat and glandular organs could be affected by the neoplasia. Soft tissue localizations have been described in subcutaneous fat of arms, legs or back, but no cases has yet been described in deep soft tissue.
Rarely, microcystic/reticular schwannoma had been described also in brain tissue, spinal cord and in the head and neck region [2-5] (Tab. I).
The rarity and variability of presentations could create problems of differential diagnosis both with mucinous-producing carcinomas (especially on small biopsies) or with other soft tissue tumors, with myxoid or reticular structure.
In the current case, the site of presentation (retroperitoneal tissue) and the lack of a diffuse myxoid stroma helped us excluding malignant soft tissue neoplasms as myxoid liposarcoma and myxofibrosarcoma, both usually with a superficial origin (mainly thigh and extremities). Morphology, with the absence of lipoblast-like elements and lacking of “chicken-wire” vessels, and MDM-2 negativity ruled out also the possibility of a dedifferentiated liposarcoma of retroperitoneum.
The intense staining with S100 and the peripheral weaker staining for EMA made us think about a nervous origin of the tumor.
Reticular perineuroma, another possibility based on morphology, usually contains slender, elongated spindle cells with bipolar cytoplasmic processes highlighted by EMA staining, and tumor cells are negative for both S-100 protein and GFAP; our case showed an opposite phenotype (EMA negative, S100+ and GFAP+).

Table I. Sites and characteristics of all described cases.

Case n°	Age/sex	Site	Growth	Reference
Digestive tract				
1	39/F	Esophagus	Unencapsulated	Gu et al. [9]
2	72/F	Stomach	Unencapsulated	Liegl et al. [1]
3	63/F	Stomach	Unencapsulated	Chetty et al. [13]
4	67/F	Mid-jejunum	Unavailable	Agaimy et al. [14]
5	93/F	Jejunum	Unencapsulated	Liegl et al. [1]
6	78/M	Small intestine	Focal infiltration	Liegl et al. [1]
7	43/F	Meso-appendix	Encapsulated	Tang et al. [10]
8	68/M	Cecum	Focal infiltration	Liegl et al. [1]
9	67/F	Cecum	Focal infiltration	Agaimy et al. [14]
10	32/F	Ascending colon	Focal infiltration	Lee et al. [15]
11	70/F	Sigmoid colon	Unencapsulated	Kienemund et al. [16]
12	70/F	Sigmoid colon	Unavailable	Kienemund et al. [16]
13	61/M	Sigmoid colon	Unencapsulated	Trivedi et al. [10]
14	73/F	Rectum	Unencapsulated	Liegl et al. [1]
Subcutaneous and soft tissue				
15	50/F	Right arm	Encapsulated	Liegl et al. [1]
16	55/M	Right forearm	Encapsulated	Chaurasia et al. [11]
17	30/F	Upper arm	Partially encapsulated	Luzar et al. [17]
18	55/M	Right upper arm	Partially encapsulated	Luzar et al. [17]
19	56/F	Back	Encapsulated	Liegl et al. [1]
20	11/M	Upper back	Unencapsulated	Liegl et al. [1]
21	28/M	Back	Partially encapsulated	Luzar et al. [17]
22	26/M	Left masticator space	Unencapsulated	Lau et al. [6]
23	69/M	Retroperitoneum	Encapsulated	Our case
Glands				
24	62/M	Pancreas	Unencapsulated	Liegl et al. [1]
25	41/M	Pancreas	Unencapsulated	Shen et al. [18]
26	53/M	Left adrenal gland	Focal infiltration	Liegl et al. [1]
27	31/F	Adrenal gland	Encapsulated	Zhou et al. [8]
28	60/M	Adrenal gland	Unencapsulated	Xie et al. [19]
29	59/F	Parotid gland	Unencapsulated	Pang et al. [20]
30	34/M	Submandibular gland	Unencapsulated	Lau et al. [6]
Others				
31	76/F	Bronchus	Unencapsulated	Liegl et al. [1]
32	22/F	Frontal lobe	Unencapsulated	Pearson et al. [2]
33	61/F	Right mandible	Focal infiltration	Yin et al. [7]
34	28/M	Right neck	Encapsulated	Gong et al. [21]
35	22/F	Palate	Encapsulated	Guo et al. [22]
36	35/M	Cervical spine	Unencapsulated	Li et al. [23]
37	40/M	Lumbar spinal canal	Encapsulated	Liu et al. [5]

The possibility of a carcinoma with mucinous production and melanoma were both excluded on morphology and with immunohistochemical reactions.

The most striking feature of our case was the high number of vascular structures admixed with spindle cells: stainings with CD31 and CD34 helped us to demonstrate a non-hyalinized wall and a regular course of vessels, without an alteration in their structure. This let us excluded also vascular neoplasms, such as hemangioma or angiosarcoma.

Conclusions

The case we present is, to the best of our knowledge, the first described in the retroperitoneum, a site where the exclusion of other mesenchymal malignancies is mandatory, united with, in the specific patient, the exclusion of a lymphoproliferative disorder.
We hope that this description will help add information about this rare and unusual neoplasm.

Conflict of interest

The Authors declare no conflict of interest.

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Ethical consideration

Neither patient consent nor ethical approval was sought: all patients attending San Martino Hospital are informed about the use of their data for continuing research at the Hospital.

Authors' contribution

RB, GG: conception, data collection, drafting and final review. GFO, JLR: review and corrections. BS, VGV: drafting and corrections.

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