Revista Española de Patología 58 (2025) 100791

SEAP-IAP

REVISTA ESPAÑOLA DE Patología Spanish Journal of Pathology

www.elsevier.es/patologia

BRIEF REPORT

Axillary intranodal palisaded myofibroblastoma, a rare tumour at an unusual site, with literature review

Durre Aden, Minnat Sharma, Sufian Zaheer*, Sunil Ranga

VMMC and Safdarjang Hospital, New Delhi, India

Received 30 April 2024; accepted 9 July 2024

KEYWORDS
Intranodal palisaded myofibroblastoma;
IPM;
Benign tumour;
Lymph node

Abstract Intranodal palisaded myofibroblastoma (IPM) arising in the axilla is an extremely rare, benign mesenchymal tumour. It is believed to originate from myofibroblast or smooth muscle cells and exhibits specific histopathological features. While there have been occasional cases of recurrence, no malignant transformation has been observed. We describe the case of a 35-year-old male presenting with an axillary mass. Histopathology revealed a tumour with a pseudo-capsule that contains compressed lymphoid tissue with spindle cells arranged in a palisade-like pattern and extravasation of red blood cells within the spindle cells. Additionally, amianthoid fibres and fuchsinophilic bodies are present. Immunohistochemical analysis typically was positive for SMA and cyclin D1, with a low proliferative index (Ki67 of <1%). The diagnosis was intranodal palisading myofibroblastoma. Only two cases of IPM in the axilla have been previously reported. Pathologists should keep this rare entity with characteristic histopathological findings in mind when reporting such tumours at an unusual site.

PALABRAS CLAVE
Miofibroblastoma en empalizada intraganglionar;
MIP;
Tumor benigno;
Ganglio linfático

Miofibroblastoma axilar intraganglionar en empalizada, un tumor raro en una localización inusual, con revisión de la literatura

Resumen El miofibroblastoma en empalizada intraganglionar (MIP) que surge en la axila es un tumor mesenquimal benigno extremadamente raro. Se cree que se origina a partir de miofibroblastos o células musculares lisas y tiene hallazgos histopatológicos específicos. Si bien ha habido casos ocasionales de recurrencia, no se ha observado una transformación maligna. Describimos el caso de un varón de 35 años que presenta una masa axilar. La histopatología reveló un tumor con una pseudocápsula que contiene tejido linfoide comprimido con células fusiformes dispuestas en un patrón similar a una empalizada y extravasación de glóbulos rojos dentro de las células fusiformes. Además, están presentes fibras amiantoides y cuerpos

Abbreviations: IPM, intranodal palisaded myofibroblastoma; IHC, immunohistochemistry; IMT, inflammatory myofibroblastic tumour; SMA, smooth muscle actin.

* Corresponding author.
E-mail address: sufianzaheer@gmail.com (S. Zaheer).

https://doi.org/10.1016/j.patol.2024.100791

fucsinófilos. El análisis inmunohistoquímico fue típicamente positivo para AME y ciclina D1, con un índice proliferativo bajo (Ki67 < 1%). Se diagnosticó miofibroblastoma en empalizada intraganglionar. Solo hay 2 casos de MIP en la axila que se han comunicado previamente. Los patólogos deben tener en cuenta esta rara entidad con hallazgos histopatológicos característicos al informar de dichos tumores en un sitio inusual.

Introduction

Intranodal palisaded myofibroblastoma (IPM) is a rare, painless, slow-growing tumour of the lymph nodes. The overall mean age at the time of diagnosis of IPM is usually 45–55 years with a male preponderance of 2:1.[1,2] Initially, it was misdiagnosed as a benign mesenchymal lesion such as neurilemmoma, schwannoma or leiomyoma occurring inside the lymph node. However, Weiss et al. identified it as a distinct entity and named it ''palisaded myofibroblastoma''.[3] The term ''intranodal'' was later added, and now it is called intranodal palisading myofibroblastoma (IPM).[4] It is also known as solitary spindle cell tumour with myoid differentiation of the lymph node or intra-nodal haemorrhagic spindle cell tumour with amianthoid fibres.[5–7] There are few cases of IPM reported in the literature, occurring in cervical lymph nodes, submandibular or inguinal nodes, although only two cases reported in axillary lymph nodes. The lesion is usually asymptomatic, which is why most patients do not seek medical attention. Gradually, the mass becomes painful and enlarges,[4,8] sometimes leading to a diagnostic dilemma for both clinicians and pathologists. Histopathology and immunohistochemistry help in the confirmatory diagnosis of such lesions. Here we present a rare case of IPM arising from the axilla, emphasizing its inclusion in the differential diagnosis of mesenchymal tumours in the axillary region.

Case report

A 35-year-old man presented with a painless, slow-growing mass in the right axilla that had been present for one year. The mass was firm, well-defined and measured 4 cm × 3 cm in diameter. Ultrasonography of the right axilla showed a solid mass with mixed echogenicity and lobulated margins, measuring 4.2 cm × 3.2 cm, raising suspicion of a neoplastic aetiology. A malignant lesion was suspected and excision biopsy was advised for confirmation. Surgical excision was performed and the specimen was sent to the histopathology department. Gross examination revealed a nodular and well-defined tumour surrounded by a pseudo-capsule, measuring 4 cm × 3.2 cm × 2 cm × 3 cm. On the cut section, the tumour was grey-white with a whorled appearance. Subcapsular haemorrhage was observed, resembling ''milk freshly poured into tea''. Multiple sections were taken and further processed for microscopic examination. Microscopy revealed an intra-nodal mass compressed by a thick fibrocollagenous capsule with remnants of lymphoid tissue at the periphery (Fig. 1a). The tumour showed uniform fascicles of spindle-shaped cells with abundant cytoplasm. Focal palisading patterns were observed, along with eosinophilic amianthoid fibres characterized by a central eosinophilic core surrounded by a paler eosinophilic, stellate-shaped periphery (Fig. 1b). Intraparenchymal haemorrhage and hemosiderin-laden macrophages were also observed among the spindle cells (Fig. 1c). There was no evidence of necrosis, nuclear atypia, or mitosis. The tumour cells were positive for smooth muscle actin (SMA) and exhibited characteristic nuclear cyclin D1 expression (Fig. 2a and b). They were negative for S100 protein, HMB-45, GFAP, CD31, CD34, CD117, Desmin, EMA, and cytokeratin. Less than 1% of tumour cells were positive for Ki67.

Discussion

IPM is an uncommon benign neoplasm characterised by spindle cell proliferation and smooth muscle differentiation.[9,10] To date, approximately 100 cases of IPM have been reported in inguinal lymph nodes, three cases in the submandibular LN, two cases each in axillary, mediastinal, retroperitoneal, and cervical LN, and one case in the parotid region LN[11] (Table 1). However, IPM occurring in axillary lymph nodes is extremely unusual.[11]

Due to the rarity of this entity, the precise aetiology and pathogenesis of IPM are not yet known. The majority of cases arise in the inguinal lymph nodes, possibly because of an increased number of smooth muscle cells and myofibroblasts, secondary to rich vascular channels and increased drainage function. This is supported by the observation that myofibroblasts in the inguinal region show positive staining for SMA and vimentin and negative staining for desmin in myofibroblasts in the inguinal region compared to fibroblasts elsewhere in the body. However, the occurrence of IPM in lymph nodes in other parts of the body contradicts this hypothesis. Some have proposed that unspecified mutagenic elements, potentially spurred by trauma, vascular stasis, or inflammation impacting the inguinal lymph nodes, could play a role in the development of the tumour.[12]

A viral aetiology has also been suggested. Recently, increased expression of cyclin D1 has been suggested in the oncogenesis; however, its positivity is seen in 50% of spindle cell tumors.[8,10] In a study by Laskin et al., it was proposed

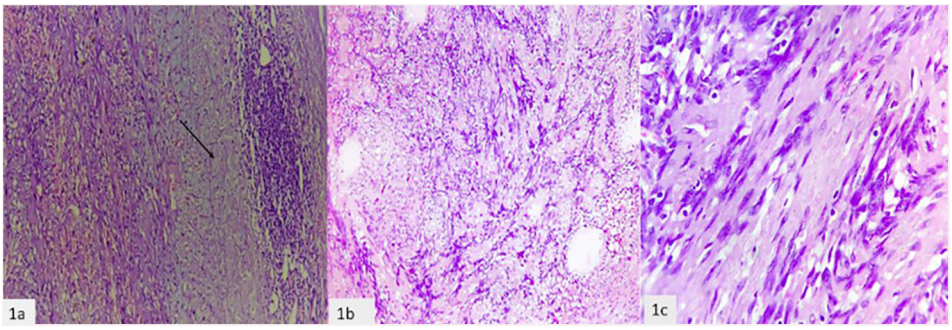

Figure 1 (a) Photomicrograph shows remnants of lymphoid compressed by a collagenous capsule (arrow) with the presence of characteristic subcapsular haemorrhage (H&E stain, 10×). (b) Photomicrograph shows features of nuclear palisading, the amianthoid-like fibre of IPM having a stellate shape with a deeply eosinophilic centre and paler periphery b (H&E stain, 10×). (c) Photomicrograph shows cellular proliferation of spindle-shaped cells arranged in loose whorls and fascicles with scant, fibrillary, eosinophilic cytoplasm, and elongated cytologically bland nuclei (H&E stain, 40×).

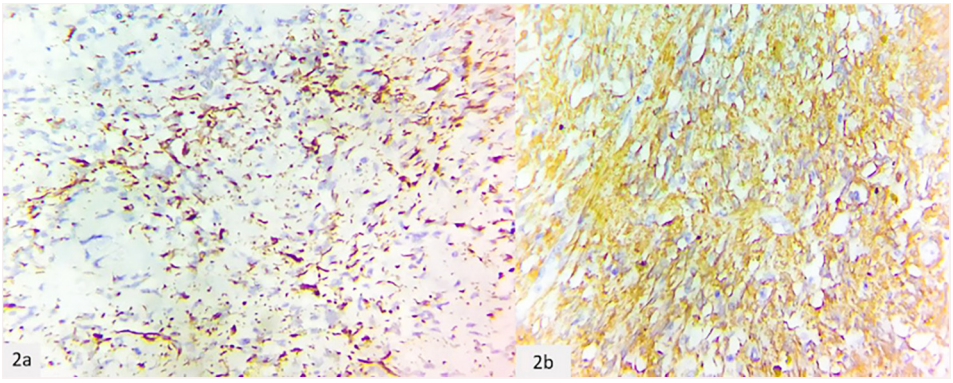

Figure 2 (a) Tumour cells are positive for cyclin D1 on immunohistochemistry (40×). (b) Tumour cells are positive for smooth muscle actin (SMA) on immunohistochemistry (10×).

that mutational activation of the β-catenin gene (CTNNB1) is likely a pivotal event in the pathogenesis of IPM.[13]

To identify these tumours, it is important to obtain a clinical history of the primary tumour and an appropriate immunohistochemical profile. Grossly the cut surface of the lymph node appears firm, grey-white in colour, with irregular haemorrhagic areas. Microscopically, features characteristic of IPM are observed, including fascicles of spindle cells with nuclear palisading, amianthoid fibres, hemosiderin pigment, and extravasated erythrocytes. There is no nuclear atypia, and mitoses are rare. Immunohistochemical staining shows that the spindle cells are positive for SMA and vimentin.[10]

As IPM is composed of spindle cells, the most important histological differential diagnoses include schwannoma, Kaposi's sarcoma, pseudomyogenic haemangioendothelioma, inflammatory myofibroblastic tumour, and primary and metastatic malignant tumours.[14,15] An essential step to distinguish IPM from non-IPM tumours or pseudotumours involves differentiating clinically, histomorphologically, and immunohistochemically. Spindle cells with nuclear palisading may be consistent with schwannoma; however, it is very unlikely to occur inside the lymph node since lymph nodes lack peripheral nerve innervation. Immunohistochemically, schwannoma shows S100 protein positivity, which is not seen in IPM. Malignant schwannoma additionally shows a raised Ki67 index. Spindle cell melanoma can be excluded by negative HMB-45 and S100 protein by IHC.[7,9,10]

Extravasation of erythrocytes and hemosiderin pigment may be suggestive of Kaposi sarcoma, and fuchsinophilic bodies of IPM can resemble hyaline globules in Kaposi sarcoma. Amianthoid fibres and nuclear palisading are not usually observed in KS. However, it shows positivity for HHV-8 (LANA1). Moreover, the absence of immunoreactivity with endothelial markers also supports the diagnosis of IPM.[6,8] Spindle cell proliferation with extensive haemorrhage raises the possibility of pseudomyogenic haemangioendothelioma, which can occur in lymph nodes. However, it will test positive for CD31, CD34, and factor 8.

Inflammatory myofibroblastic tumour (IMT) shows spindle cell proliferation in the lymph node, with compressed endothelium forming vascular structures and a dense inflammatory infiltrate. The absence of amianthoid fibres and a plasma cell-rich inflammatory infiltrate are important features that help distinguish this entity from IPM.[3] On immunohistochemistry (IHC) it shows positivity for ALK.

The treatment for IPM is surgical excision.[10] IPM has a good prognosis with a very low recurrence rate.[9] Therefore, it is crucial to distinguish it from metastatic lesions of the lymph node with spindle cell morphology, such as metastases from spindle cell melanoma, carcinoma with

Table 1 Non-inguinal region, cases of IPM.

Author and year	Site	Age (Yr.)/sex	Size (cm)	Treatment	Outcome	Follow-up
Sagar et al., 2011[8]	Right side, retroperitoneum	72/M	8 × 8	Excision	No recurrence	7 years
Bouhajja et al., 2017[10]	Right side, submandibular	44/F	3 × 2	Excision	Well	5 years
Fletcher et al., 1990[17]	Right submandibular	40/F	1.5	Excision	Well	2½ years
Alguacil-Garcia, 1992[16]	Right submandibular	61/M	2.6 × 1.9	Excision	–	–
Michal et al., 1993[18]	Cervical neck region	F		Excision	–	–
Bhullar et al., 2013[19]	Right axilla	39/F	6 × 5.5	Excision	Well	10 months
D'Antonio et al., 2014[20]	Right axilla	25/F	3 × 3	Excision	Well	2 years
Laskin et al., 2015[13]	Left cervical neck mass	38/M	–	Excision	–	–
Hicham et al., 2015[21]	Right retroperitoneum	39/M	2 × 1.5	Excision	Discharged without complications	–
Yim et al., 2016[22]	Mediastinum, right paratracheal	68/M	5.9 × 3.6	Video-assisted thoracic surgical posterior approach excision	The patient had associated adenocarcinoma, right upper lobe lung	–
Fatani et al., 2018[23]	Right parotid region	07/M	4.5	Excision	–	–
Hu et al., 2019[24]	Mediastinum, right para-oesophageal	70/F	5 × 3.1	Robotic-assisted posterior mediastinal mass excision	–	–

pseudo-sarcomatous features, and sarcoma. Metastasis to the lymph node can be diagnosed by the clinical history of the primary tumour, pleomorphism, marked atypia, high mitotic activity, and specific immunohistochemical characteristics.[16]

Conclusion

IPM is a rare benign lesion that can arise in the inguinal, mandibular, and cervical lymph nodes. It can rarely be seen in the axillary lymph node. Immunohistochemistry plays a crucial role in ruling out other differential diagnoses. An early and timely biopsy allows for effective and early patient care and reduces the risk of disease recurrence.

CRediT authorship contribution statement

Dr Durre Aden: concepts, design, definition of intellectual content, literature search, data acquisition, manuscript preparation, manuscript editing, and manuscript review.

Minnat Sharma: diagnosis, preparation of the manuscript, and search for references.

Dr Sufian Zaheer: manuscript preparation, literature search, manuscript editing, and manuscript review.

Dr Sunil Ranga: overall manuscript preparation, manuscript editing, and manuscript review.

Informed consent

It is a retrospective case report and consent was obtained from the patient. The study was performed according to Declaration of Helsinki guidelines.

Funding

The authors declare that no financial support was received.

Conflict of interest

The authors declare that they have no conflict of interest.

References

1. Ioachim HL, Medeiros LJ. Spindle cell neoplasms of the lymph nodes. Lymph node pathology. Philadelphia: JB Lippincott Publishers Company; 2008. p. 571–3. ISBN-13: 978-0781775960.
2. Margaret A-K, Wright P, Wright D. Non-hematolymphoid tumours of the lymph node. Diagnostic lymph node pathology. FL: CRC

Press, Taylor & Francis Group; 2016. p. 163–4. ISBN 13: 978-1-4987-3270-3.
3. Weiss SW, Gnepp DR, Bratthauer GL. Palisaded myofibroblastoma. A benign mesenchymal tumor of lymph node. Am J Surg Pathol. 1989;13:341–6 [PMID: 2712186].
4. Nguyen T, Eltorky MA. Intranodal palisaded myofibroblastoma. Arch Pathol Lab Med. 2007;131:306–10, http://dx.doi.org/10.5858/2007-131-306-IPM.
5. Nsour E, Al Khader A. A rare case of intranodal hemorrhagic spindle cell tumor with amianthoid fibers presenting as a suspicious lump in the groin. Int J Surg Case Rep. 2019;64:72–4, http://dx.doi.org/10.1016/j.ijscr.2019.09.042.
6. Kandemir NO, Barut F, Ekinci T, Karagülle C, Ozdamar SO. Intranodal palisaded myofibroblastoma (intranodal hemorrhagic spindle cell tumor with amianthoid fibers): a case report and literature review. Diagn Pathol. 2010;5:12, http://dx.doi.org/10.1186/1746-1596-5-12.
7. Lee JY, Abell E, Shevechik GJ. Solitary spindle cell tumour with myoid differentiation of the lymph node. Arch Pathol Lab Med. 1989;113:547–50.
8. Sagar J, Vargiamidou A, Manikkapurath H. Intranodal palisaded myofibroblastoma originating from retroperitoneum: an unusual origin. BMC Clin Pathol. 2011;11:7, http://dx.doi.org/10.1186/1472-6890-11-7.
9. Karabulut YY. Pathological approach to a special benign tumor with regard to its differential diagnose spectrum: intranodal palisaded myofibroblastoma. J Immunol Sci. 2018;25:13–6.
10. Bouhajja L, Jouini R, Khayat O, Koubâa W, Mbarek C, Ben Brahim E, et al. Intranodal palisaded myofibroblastoma in a submandibular lymph node. Case Rep Otolaryngol. 2017;2017:7121485, http://dx.doi.org/10.1155/2017/7121485.
11. Creager AJ, Garwacki CP. Recurrent intranodal palisaded myofibroblastoma with metaplastic bone formation. Arch Pathol Lab Med. 1999;123:433–6, http://dx.doi.org/10.5858/1999-123-0433-RIP.
12. Barbareschi M, Mariscotti C, Ferrero S, Pignatiello U. Intranodal haemorrhagic spindle cell tumour: a benign Kaposi-like nodal tumour. Histopathology. 1990;17:93–6, http://dx.doi.org/10.1111/j.1365-2559.1990.tb00672.x.
13. Laskin WB, Lasota JP, Fetsch JF, Felisiak-Golabek A, Wang ZF, Miettinen M. Intranodal palisaded myofibroblastoma: another mesenchymal neoplasm with CTNNB1 (β-catenin gene) mutations: clinicopathologic, immunohistochemical, and molecular genetic study of 18 cases. Am J Surg Pathol. 2015;39:197–205, http://dx.doi.org/10.1097/PAS.0000000000000299.
14. Haddad A, Marwan K. Intranodal palisaded myofibroblastoma: a diagnostic differential for inguinal lymphadenopathy. Am J Case Rep. 2021;22:e934752, http://dx.doi.org/10.12659/AJCR.934752.
15. Pena-Burgos EM, Tapia-Viñe M, Rodríguez-García AM, Ortiz-Cruz EJ, Pozo-Kreilinger JJ. Cytopathological findings of intranodal palisaded myofibroblastoma: case report and review of the literature. Diagn Cytopathol. 2023;51:E248–54, http://dx.doi.org/10.1002/dc.25172.
16. Alguacil-Garcia A. Intranodal myofibroblastoma in a submandibular lymph node. A case report. Am J Clin Pathol. 1992;97:69–72, http://dx.doi.org/10.1093/ajcp/97.1.69.
17. Fletcher CD, Stirling RW. Intranodal myofibroblastoma presenting in the submandibular region: evidence of a broader clinical and histological spectrum. Histopathology. 1990;16:287–93, http://dx.doi.org/10.1111/j.1365-2559.1990.tb01117.x.
18. Michal M, Chlumská A, Skálová A, Fakan F. Palisaded intranodal myofibroblastoma. Electron microscopic study. Zentralbl Pathol. 1993;139:81–8.
19. Bhullar JS, Varshney N, Dubay L. Intranodal palisaded myofibroblastoma: a review of the literature. Int J Surg Pathol. 2013;21:337–41, http://dx.doi.org/10.1177/1066896913489348 [Epub 27.05.13; PMID: 23714684].
20. D'Antonio A, Addesso M, Amico P, Fragetta F. Axillary intranodal palisaded myofibroblastoma: report of a case associated with chronic mastitis. BMJ Case Rep. 2014;2014, http://dx.doi.org/10.1136/bcr-2014-205877, bcr2014205877.
21. Hicham L, Khmou M, Najih M, Zouaidia F, Sair K, El Khannoussi B. Intranodal palisaded myofibroblastoma: a case report from an unusual site. Pan Afr Med J. 2015;22:78, http://dx.doi.org/10.11604/pamj.2015.22.78.7884.
22. Yim IH, Will MB, Dhaliwal C, Salter DM, Walker WS. Intranodal palisaded myofibroblastoma masquerading as N2 non-small cell lung carcinoma. Ann Thorac Surg. 2016;102:e47–8, http://dx.doi.org/10.1016/j.athoracsur.2015.11.060.
23. Fatani H, Alsamman A, AlJabab A, Al Dandan S. Intranodal palisaded myofibroblastoma arising from parotid gland: a case report of an unusual origin. Hum Pathol Case Rep. 2018;13:16–7, http://dx.doi.org/10.1016/j.ehpc.2018.03.007.
24. Hu J, Tipps AMP, Hasteh F, Lin G, Zare SY. Intranodal palisaded myofibroblastoma of para-esophageal lymph node: case report with cytologic and histologic findings. Diagn Cytopathol. 2019;47:1306–9, http://dx.doi.org/10.1002/dc.24303.