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Case Report

Surgery Section

Follicular Dendritic Cell Sarcoma with Nodal Involvement in A Young Woman: A Case Report

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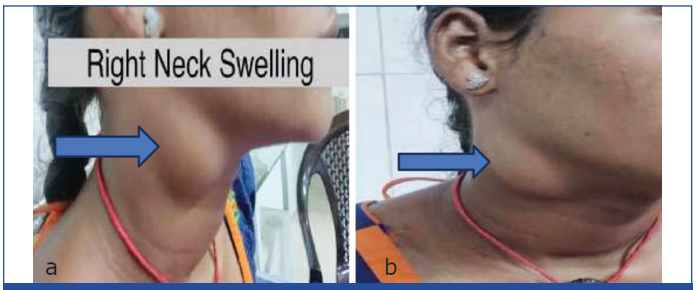
ABSTRACT

Follicular Dendritic Cell Sarcoma (FDCS) is a rare tumour that develops from follicular dendritic cells, which are essential to the immune system. It is more frequently seen in adults, with lymph node involvement being common, while extra-nodal occurrences are rare. This case report describes a 17-year-old female who presented with a four-year history of a progressively enlarging, painless swelling on the right side of the neck. Clinical examination indicated a soft, fluctuant, mobile, non-tender mass situated below the right mandibular angle, without regional lymphadenopathy. Imaging studies, including Magnetic Resonance Imaging (MRI) and cerebral angiography, identified a well-defined lesion in the right submandibular region with heterogeneous hyperintensity and internal cystic changes, abutting the salivary gland and carotid vessels, but without vascular invasion. Fine Needle Aspiration Cytology (FNAC) suggested differential diagnoses of paraganglioma and lymphoproliferative disorder. The patient underwent a wide local excision and biopsy under general anaesthesia. Intraoperatively, the tumour was well-encapsulated but near critical vascular structures, necessitating careful dissection. Histopathological and Immunohistochemical (IHC) evaluation confirmed the diagnosis of FDCS, a rare low-grade malignancy of histiocytic/dendritic cell origin, with nodal involvement. Postoperative Positron Emission Tomography-Computed Tomography (PET-CT) revealed no evidence of residual or metastatic disease. The patient's recovery was uneventful, and no recurrence was detected during follow-up.

Keywords: Cervical lymphadenopathy, Immunohistochemistry, Metastatic disease, Submandibular region

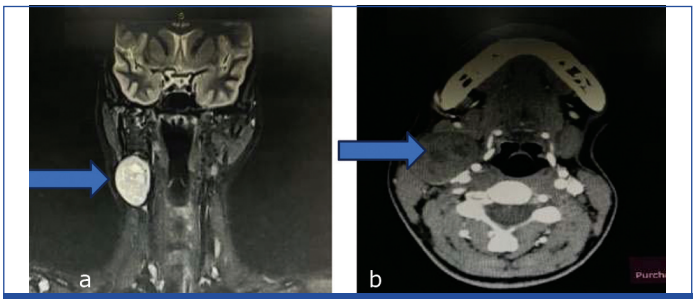
CASE REPORT

A 17-year-old female presented with a gradually enlarging, painless swelling on the right side of her neck [Table/Fig-1a,b]. The swelling had been progressively increasing in size over the past four years. It measured approximately 5×3 cm and was located just below the right mandibular angle. On examination, the mass was soft, mobile, fluctuant, and non-tender, with normal overlying skin and no regional lymphadenopathy. There were no associated symptoms such as trauma, discharge, fever, weight loss, fatigue, or signs suggestive of endocrine hyperfunction like tremors or palpitations. The patient denied any dysphagia, dyspnea, or changes in voice. Her medical, surgical, and family histories were unremarkable. On general examination, she was alert, oriented, and haemodynamically stable with a blood pressure of 110/70 mmHg and a pulse rate of 80 beats per minute.

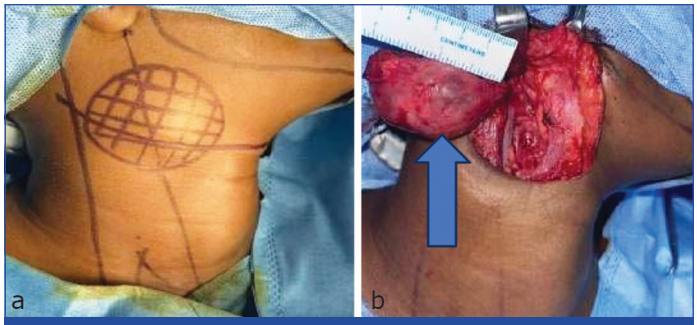


[Table/Fig-1a,b]: Clinical presentation showing a right-sided neck swelling located just below the angle of the mandible.

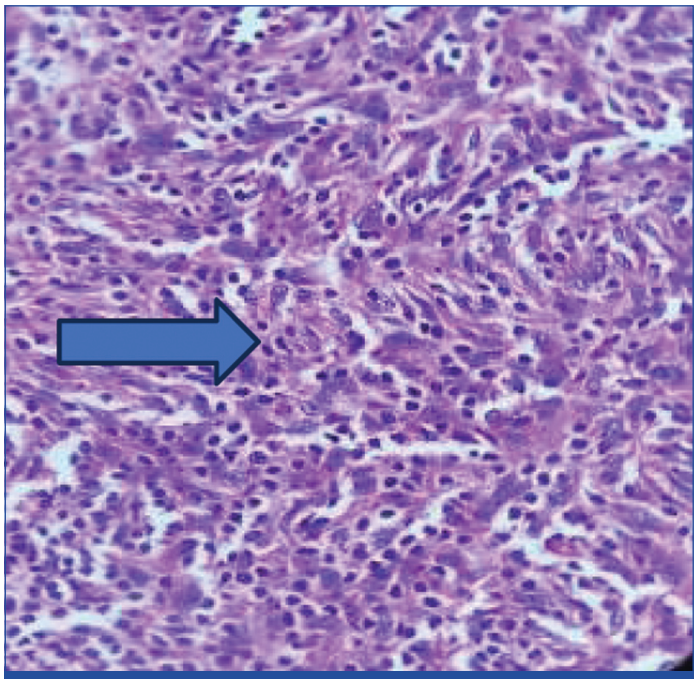
Magnetic Resonance Imaging (MRI) of the neck revealed a well-defined, heterogeneously hyperintense lesion on T2-weighted images with internal cystic areas in the right submandibular region. The lesion was adjacent to the submandibular gland and carotid vessels, extending from the C2 to C4 vertebral level, causing mild medial displacement without any signs of invasion [Table/Fig-2a]. Cerebral angiography showed a heterodense lesion with mild enhancement located near the right facial artery and carotid arteries, without evidence of vascular encroachment [Table/Fig-2b]. FNAC of the mass revealed a population of round to polygonal and spindle-shaped plasmacytoid cells arranged in rosette and acinar patterns. Based on these findings, the initial differential diagnoses included paraganglioma and lymphoproliferative disorder. The patient underwent a wide local excision and biopsy under general anaesthesia. Intraoperatively, the mass appeared well-encapsulated but was closely adherent to the carotid vessels. Complete excision was performed with preservation of surrounding vital structures, and the specimen was sent for histopathological examination [Table/Fig-3a,b]. Histopathological evaluation revealed a well-encapsulated tumour composed of spindle-shaped and ovoid cells arranged in fascicles and storiform patterns, with scattered multinucleated and binucleated tumour cells [Table/Fig-4]. IHC staining confirmed the diagnosis of FDCS, a rare low-grade sarcoma of histiocytic/dendritic origin, with evidence of nodal involvement [Table/Fig-5a-g,6]. Following surgical excision, a postoperative PET-CT scan was performed and showed no evidence of hypermetabolic residual disease or distant metastasis. The surgery was technically challenging due to the tumour's proximity to major vascular structures and the submandibular gland, requiring careful dissection and meticulous intraoperative navigation to avoid vascular injury. The patient had an uneventful postoperative recovery, and follow-up at 7, 30, and 90 days, evaluations revealed no signs of recurrence.



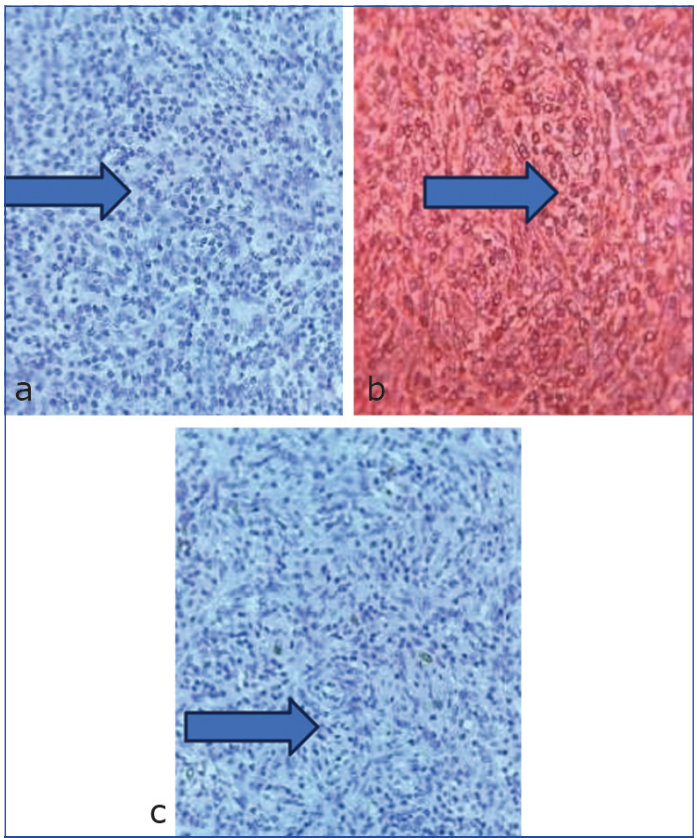
[Table/Fig-2]: MRI (a) showing a well-defined heterogeneously hyperintense lesion with internal cystic components in the right submandibular region. Cerebral angiography; b) showing a heterodense mass near the carotid vessels without vascular invasion.



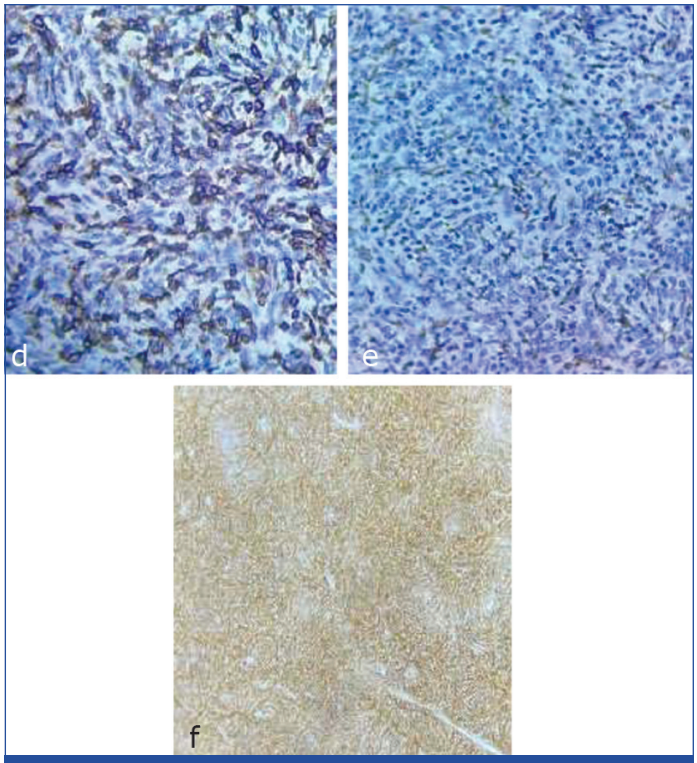
[Table/Fig-3a,b]: Intraoperative findings depicting the encapsulated mass closely abutting the carotid vessels and the gross surgical specimen. Storiform pattern observed.



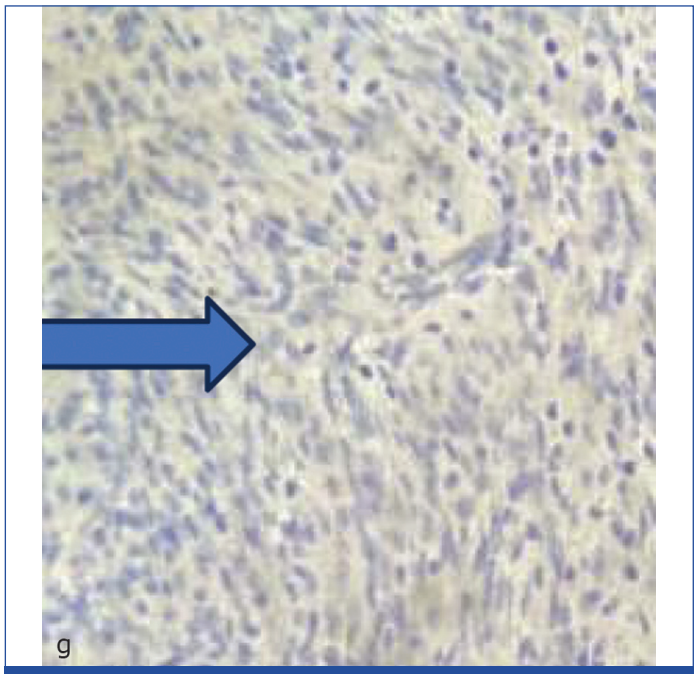
[Table/Fig-4]: Histopathological image showing spindle and ovoid cells in storiform and fascicular patterns; Vimentin-positive staining, observed at 10x magnification.



[Table/Fig-5a-c]: Immunohistochemical (IHC) positivity in tumour cells for CD34,Vimentin, and CD68, observed at 10x magnification.



[Table/Fig-5d-f]: Negative immunoreactivity for CD45 and anaplastic lymphoma kinase (ALK) in tumour cells, consistent with dendritic cell origin, observed at 10x magnification.



[Table/Fig-5g]: Positive CD21 and CD23 staining confirming follicular dendritic cell lineage, observed at 10X magnification.

Positive tumour marker	Negative tumour marker
Vimentin	CD34
CD68	CD45
CD21	ALK
CD23	
Lymphoid cells	
CD45	

[Table/Fig-6]: Immunohistochemistry summary table of marker expression in tumour and lymphoid cells.

DISCUSSION

The FDCS is a rare malignant tumour originating from follicular dendritic cells, which are crucial for antigen presentation and B-cell regulation within lymphoid tissues. It accounts for less than 0.5% of

all soft tissue sarcomas. Although it generally occurs in adults, the age range is broad, and there is a slight female predominance. [1] Patients typically present with a gradually enlarging, painless mass, most often in cervical or mediastinal lymph nodes, though extra nodal sites such as the liver, spleen, and gastrointestinal tract may also be involved [1]. The current case stands out due to the patient's young age (17 years) and nodal localisation without systemic features. Initial imaging and FNAC were non-specific, reflecting the diagnostic complexity of FDCS. As supported by Wang H et al., IHC staining remains essential, with CD21 and CD35 being the most reliable markers [2]. Previous literature reported more aggressive presentations with widespread bone and abdominal involvement, often requiring chemotherapy due to inoperability [1]. In contrast, our case was amenable to complete surgical resection, with no postoperative residual disease, consistent with outcomes seen in cases managed surgically at an early stage [3,4]. Recent studies have noted Programmed Death-Ligand 1 (PD-L1) expression in some FDCS cases, suggesting a potential future role for immunotherapy [4,5]. However, standardised treatment protocols are lacking due to their rarity. Screening is not currently recommended, but persistent lymphadenopathy, especially with atypical imaging or inconclusive cytology, should raise suspicion. This case highlights the value of early surgical intervention in localised FDCS and reinforces the need for long-term surveillance due to potential recurrence. Broader data collection and reporting will be a key to refining diagnostic and treatment guidelines for this rare tumour.

CONCLUSION(S)

The FDCS is an uncommon and often misdiagnosed malignancy. Its occurrence in adolescents is exceptionally rare, necessitating a thorough histopathological and IHC evaluation for accurate diagnosis. Surgical excision remains the treatment of choice, with close postoperative monitoring required to detect recurrence. This case underscores the importance of considering FDCS in persistent cervical lymphadenopathy cases.

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