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RARE OPHTHALMIC PRESENTATIONS OF SPINDLE CELL SARCOMA: A CASE SERIES FROM EASTERN INDIA

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ABSTRACT **Purpose:** To report six cases of spindle cell sarcoma (SCS) with rare ophthalmic presentations, highlighting their diverse anatomical involvement, diagnostic challenges, and outcomes in a resource-limited setting. **Methods:** We retrospectively analyzed six patients with biopsy-proven spindle cell sarcoma involving ocular and periocular regions. The age ranged from 6 to 65 years. Clinical features, imaging, surgical outcomes, histopathology, and follow-up data were reviewed. **Results:** The spectrum included: (1) rapidly progressive orbital tumor in a 6-year-old, (2) basifrontal mass in a young adult with papilledema, (3) 12-year-old boy with recurrent lower lid mass, (4) 65-year-old female with conjunctival SCS, (5) 38-year-old man with sinonasal-origin SCS with proptosis, and (6) 41-year-old with relapsing ptosis and superior sulcus mass. All patients underwent surgical excision; three had adjuvant radiotherapy. Recurrence was noted in two cases within 3 months. Socioeconomic constraints affected follow-up in four patients. **Conclusion:** Ophthalmic spindle cell sarcoma, while rare, may present in diverse forms. Early imaging, prompt histopathologic diagnosis, and aggressive multidisciplinary management are essential for better outcomes. Socioeconomic factors and poor follow-up significantly affects prognosis in under-resourced regions.

KEYWORDS : Spindle Cell Sarcoma, Orbital Humeroulnar Oncology, Orbital Tumour.

INTRODUCTION

Spindle cell carcinoma (SpCC), also referred to as sarcomatoid carcinoma, is a rare and aggressive variant of squamous cell carcinoma (SCC) characterized by biphasic histopathological features—malignant epithelial components intermingled with spindle-shaped mesenchymal elements. This dual differentiation is attributed to epithelial–mesenchymal transition (EMT), contributing to the tumour's pronounced invasiveness and metastatic potential [1,2].

While SpCC predominantly involves the upper aerodigestive tract—including the oral cavity, larynx, and pharynx—it is exceedingly rare in the ophthalmic region, particularly within the orbit and ocular adnexa [3]. In reported instances, clinical presentation may be vague, manifesting as painless swelling, orbital masses, or periorbital discomfort, which often leads to diagnostic delays. Immunohistochemistry is pivotal in differentiating SpCC from other spindle cell neoplasms, typically revealing co-expression of epithelial markers (e.g., cytokeratin) and mesenchymal markers (e.g., vimentin) [4].

Only a limited number of cases involving periocular or orbital SpCC have been documented. Nair et al. described a 68-year-old female with a medial canthal mass initially misdiagnosed as atypical fibro-xanthoma, which was later confirmed as SpCC via excisional biopsy [5]. Similarly, Ahmad et al. reported a 61-year-old male with periorbital pain and blepharoptosis, where imaging and histo-pathology confirmed primary orbital SpCC [6]. These isolated cases highlight the diagnostic complexity and variable clinical behaviour of such tumours.

The prognosis for orbital or intracranial spindle cell sarcomas remains guarded due to their high recurrence rates, lymph vascular invasion, and potential for distant metastases [1,3]. Surgical excision with adjuvant therapies forms the mainstay of management, although outcomes depend heavily on early detection and comprehensive follow-up.

In this case series, we present six rare and varied ophthalmic presentations of spindle cell sarcoma—including orbital, conjunctival, and intracranial manifestations. The series spans a broad age range and highlights the diagnostic challenges, radiologic ambiguity, and impact of socioeconomic barriers on treatment outcomes. By sharing our experience in a resource-limited tertiary care setting, we aim to contribute to the growing recognition of this uncommon yet aggressive ocular malignancy.

MATERIALS AND METHODS:

This retrospective case series included six patients diagnosed with spindle cell sarcoma (SCS) involving ocular or periocular structures. The study was conducted in regional institute of ophthalmology in Eastern India during the period of March 2024 to April 2025.

Patients clinical data, ophthalmic presentation, imaging data, surgical notes, histopathological data were noted and analysed.

RESULTS

The study consisted of 6 patients with male preponderance (4 males and 2 females). The mean age was 35.1 years (range 6-65years) . Clinical presentations and anatomical involvement varied widely, reflecting the aggressive and heterogeneous nature of SCS.

Orbital Involvement (3 Cases)

Three patients presented with orbital lesions. One notable case involved a 6-year-old girl who initially presented with a small medial canthal nodule that progressively enlarged over 7 months into a large exophytic orbital mass measuring 120 × 108 mm, encasing the left globe.

Figure 1: A. Initial presentation showing a red, fleshy lesion at the medial canthus with crusting and mucoid discharge. B, C. progressive enlargement over 4 months

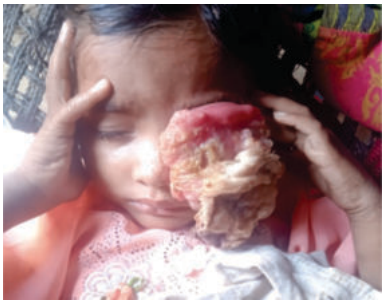

Figure 2: Large exophytic orbital mass in a 6 years old child.

MRI confirmed globe-encasing orbital extension to the apex. She was severely anaemic (Hb 5.6 g/dL), requiring transfusion before undergoing orbital exenteration with cheek advancement flap. Histopathology showed high-grade SCS with necrosis, 20–25 mitoses/HPF, pleomorphism, and lymph vascular invasion. Postoperatively, she developed graft necrosis due to poor local hygiene and socioeconomic barriers, leading to inadequate follow-up and oncology referral delay.

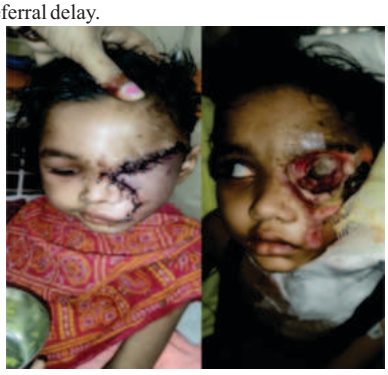

Figure 3: A. Postoperative appearance following orbital exenteration and advancement flap reconstruction. B. Postoperative image following orbital exenteration and cheek advancement flap, showing exposed wound bed with early signs of graft necrosis and poor wound hygiene.

Another orbital case was a 6-year-old boy presenting with painless proptosis and a superomedial eyelid mass. Imaging revealed a lobulated lesion displacing the globe. Biopsy confirmed SCS, and the patient was scheduled for orbitotomy and radiotherapy but surgery remained pending at the time of reporting.

The third orbital case was a 46-year-old male who presented with right eyelid swelling and ptosis. CE-MRI showed bilateral orbital involvement, including a dominant 31 × 17 mm superolateral lesion. Biopsy confirmed SCS. Systemic evaluation and surgical planning were underway.

Intracranial Involvement with Ocular Signs (1 Case): A 23-year-old male presented with bilateral visual blurring, headache, and Grade 5 papilledema. CE-MRI revealed a lobulated Basi frontal parafalcine mass extending into the nasal cavity via cribriform plate erosion. Fundus examination confirmed bilateral disc edema with peripapillary haemorrhages. He underwent extended Basi frontal craniotomy with partial tumour resection. Histopathology confirmed SCS with prominent vascularity. Despite initial recovery, early recurrence was observed within one month, highlighting the tumour's aggressive biology.

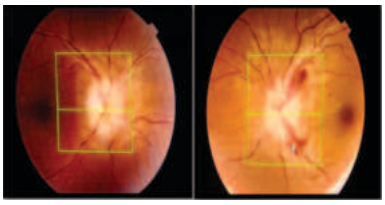

Figure 4: Fundus image showing bilateral disc edema with peripapillary hemorrhages and obscuration of vessels, consistent with Frisen grade 5 papilledema.

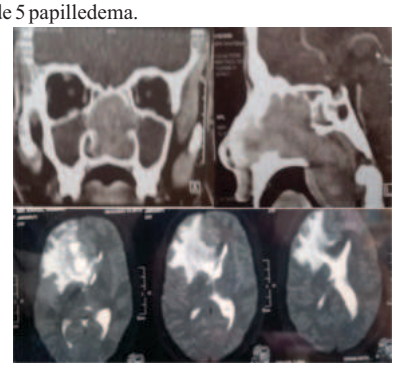

Figure 5: CE-MRI showing a right Basi frontal parafalcine mass compressing frontal lobe structures and extending through the cribriform late into the nasal cavity.

Conjunctival Presentations (2 cases):
Two 65-year-old females presented with conjunctival masses clinically mimicking ocular surface squamous neoplasia (OSSN). One had a nodular, ulcerated lesion on the upper eyelid, while the other had a fleshy temporal conjunctival mass. Both underwent excisional biopsy. Immunohistochemistry revealed cytokeratin and vimentin co-expression, confirming SCS. Neither showed orbital extension, and no recurrence was observed during a 4-month follow-up.

Management and Outcomes:
Surgical excision was planned in all cases. Three were advised adjuvant radiotherapy. Immunohistochemistry was critical in differentiating SCS from other spindle cell tumours. Two patients (Cases 1 and 2) showed early recurrence or complications, while others were either stable or awaiting definitive management. Socioeconomic factors and healthcare access limitations significantly impacted follow-up and treatment adherence in four cases

Table 1: Comparative Summary of Clinical Presentation, Imaging Findings, Histopathology, Treatment, and Outcomes in six cases of ophthalmic spindle cell sarcoma.

Case No.	Age / Gender	Presentation	Imaging Findings	Histo-pathology	Treat-ment	Outcome / Follow-up
1	6F	Progressively enlarging periocular mass with discharge and hemorrhagic spots	Large exophytic mass (120Ã—108 mm) encasing left globe extending to apex	High-grade spindle cell sarcoma with necrosis, mitosis, LVI	Orbital exenteration with cheek advancement flap	Graft necrosis; poor hygiene; referred to
2	23M	Bilateral progressive blurring of vision with papilledema and headache	Basi frontal mass compressing frontal lobe, eroding cribriform plate	Spindle cell sarcoma with prominent vascularity	Basi frontal craniotomy with partial resection	Early recurrence at 1-month post-op
3	65F	Nodular, ulcerated lesion on upper palpebral conjunctiva	No orbital extension noted	Spindle cell sarcoma (CK+, Vimentin+)	Excision biopsy with follow-up	No recurrence at 4 months
4	65F	Fleshy temporal conjunctival mass, suspicious for OSSN	No orbital extension, declined further imaging	Spindle cell sarcoma (CK+, Vimentin+)	Excision biopsy, refused further imaging	No recurrence at 4 months
5	6M	Painless proptosis with superomedial eyelid mass	Lobulated superomedial orbital mass displacing globe	Spindle cell sarcoma	Orbitotomy planned with radiotherapy	Awaiting surgery and radiotherapy
6	46M	right upper eyelid fullness and ptosis	Minimally enhancing bilateral orbital lesions; 31Ã—17 mm superolateral lesion in right orbit	Spindle cell sarcoma	Planned orbitotomy and systemic workup	Awaiting orbitotomy; systemic evaluation ongoing

DISCUSSION

Spindle cell sarcoma (SCS) is a rare, high-grade mesenchymal malignancy that presents a diagnostic challenge in ophthalmic practice due to its variable clinical appearance and resemblance to more common, benign conditions. This case series highlights six diverse ophthalmic presentations of SCS—ranging from orbital and conjunctival lesions to an intracranial tumor with ocular signs— underscoring the importance of early suspicion, biopsy, and multidisciplinary care.

Two pediatric patients presented with orbital involvement. One child developed a progressively enlarging periocular mass necessitating exenteration, with postoperative complications due to poor follow-up. The other, with a superomedial orbital mass and preserved vision, was planned for orbitotomy and radiotherapy. These cases reflect the aggressive course of orbital SCS and the consequences of delayed management, as similarly observed by Mathew et al. [7].

An unusual intracranial presentation was seen in a 23-year-old male with bilateral papilledema due to a Basi frontal mass. Despite neurosurgical intervention, early recurrence occurred, aligning with the findings of Wang et al., who reported poor outcomes in histiocytic sarcomas of the CNS despite surgical decompression [8].

Two elderly females presented with fleshy conjunctival lesions misdiagnosed clinically as ocular surface squamous neoplasia (OSSN). Excisional biopsy and immunohistochemistry revealed SCS with cytokeratin and vimentin positivity. Similar masquerading behavior has been documented by Nair et al. and Ahmad et al., emphasizing the role of histopathology in diagnosing conjunctival spindle cell neoplasms [5,6].

In one case, a 46-year-old male presented with progressive right upper eyelid swelling and mild ptosis. Imaging revealed a 31×17 mm minimally enhancing lesion in the superolateral orbit, suggestive of pseudotumor or lymphoma. However, biopsy confirmed spindle cell sarcoma. This case illustrates how even radiologically indeterminate orbital lesions may harbor aggressive pathology, reinforcing the importance of tissue diagnosis in atypical orbital masses.

Across the series, histopathological examination and immuno-histochemistry were crucial for diagnosis. Co-expression of cytokeratin and vimentin, characteristic of SCS, helped differentiate it from other spindle cell tumors and sarcomatoid carcinomas [4]. Incomplete surgical resection, socioeconomic barriers, and loss to follow-up contributed to recurrence in two cases. These findings align with earlier studies emphasizing the importance of complete excision, adjuvant therapy, and structured follow-up in improving outcomes [1–3].

CONCLUSIONS

Ophthalmic spindle cell sarcoma, though rare, can present across a wide demographic and anatomical spectrum. Its aggressive nature, potential for recurrence, and clinical overlap with benign lesions demand a high index of suspicion. Early biopsy, appropriate imaging, and multidisciplinary intervention are essential, especially in resource-limited settings where diagnostic delays can significantly affect prognosis.

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