Atypical Spindle Cell Neoplasm of the Nasal Sidewall

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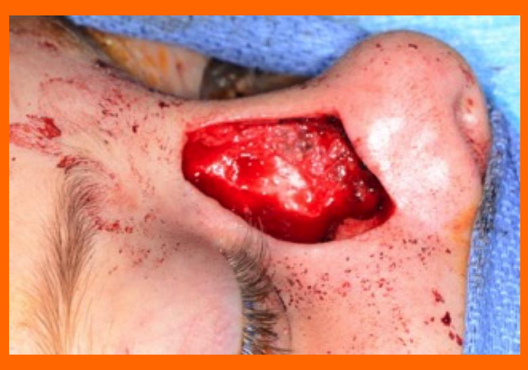
Abstract

Introduction: Spindle cells are of mesenchymal origin and characterize the morphology of a variety of benign and malignant neoplasms. Malignant spindle cell neoplasms are uncommon tumors in the head and neck region and are often described as a spindle cell variant of another common malignancy, such as squamous cell carcinoma, melanoma, or sarcomas[1,2]. In this report, we review the uncommon spindle cell neoplasms of the head and neck, report the case of a rare spindle cell neoplasm, and present a reasonable treatment plan that may assist other clinicians. The patient's malignancy is an exceedingly rare diagnosis and has not been previously described in the head and neck cancer literature.
Study Design: Case report and literature review.
Methods: The medical records of a ten year old patient with an uncommon spindle cell neoplasm of the nasal sidewall were reviewed. The PubMed database was searched for literature describing the uncommon spindle cell neoplasms of the head and neck with the key words "spindle cell" or "atypical spindle cell" with "head and neck", "nasal", "nose", or "sinus."
Results: A ten year old female patient presented with an atypical spindle cell neoplasm of the nasal sidewall that did not fit the morphological or immunohistochemical characteristics of well known diagnostic entities. The lesion recurred twice, requiring extensive nasal sidewall resection and left paramedian forehead flap reconstruction. Given the patient's age, rarity and location of the lesion, as well as psychological impact of cancer resection, decisions regarding cancer surveillance and reconstructive timing and technique were difficult and required cautious oncologic, psychological, and surgical management strategies. She continues to undergo monitoring and has had multiple revision surgeries.
Conclusions: Spindle cell neoplasms remain a diagnostic challenge and an invasive lesion may not resemble well known oncological diagnoses. For child or adolescent patients that are suffering similar conditions with a significant defect after cancer resection, we recommend a multidisciplinary approach to management, frequent contact and followup with family, and early involvement of psychology.

Case Report

A ten-year-old female presented to a private Otolaryngologist with a right nasal sidewall lesion resembling an enlarging subcutaneous acne papule that failed to resolve. Endonasal excision revealed an unclear diagnosis; the mass initially appeared to be a benign spindle cell lesion with a prominent vascular component. The pathology slides were sent for a second opinion where the neoplasm was diagnosed as an "atypical spindle cell neoplasm." Histological examination was remarkable for spindle cells with atypical tapering nuclei, an indistinct, pale cytoplasm, and locally infiltrating into skeletal muscle (Figure 1). The morphology did not fit with any well-known malignancies and the immunochemical stains for the initial specimen were negative for SMA, desmin, CD34, S100, and EMA. Upon presentation to our institution, it was noted that the lesion had recurred and a new lesion had developed adjacent to it (Figure 2). Both masses were slightly tender to palpation; however, were without overlying skin changes. CT and MRI showed a small mass in the right nasal sidewall with no obvious bony involvement.

Head and Neck tumor board recommendations included wide local excision of the nasal sidewall tumor with close follow-up. Second opinions were sought from sarcoma and pediatric oncology specialists, both of whom agreed with the tumor board's recommendations. The patient underwent resection of the soft tissue of the right nasal sidewall with the periosteum of the nasal bone as the deep margin (Figure 3). The defect was reconstructed using a split-thickness skin graft for close monitoring of recurrence. On final pathology the periosteum was positive for infiltrating spindle cell neoplasm and a new subcutaneous lesion had also developed adjacent to her prior resection site. The patient underwent resection of her second recurrence and underlying bone resection. This left a 2.5 cm x 1.5 cm through-and-through defect into the right nasal cavity (Figure 4). A nasal prosthesis was created to better camouflage the defect. After six months of monthly monitoring including nasal endoscopy the patient underwent two-stage left paramedian forehead flap reconstruction of her right-sided defect (Figure 5). She was monitored monthly for the first year following surgical resection followed by 3-month examination for the following two years. She is now re-evaluated every 4-6 months.



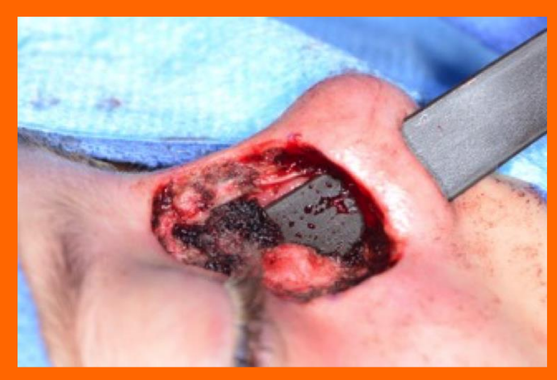
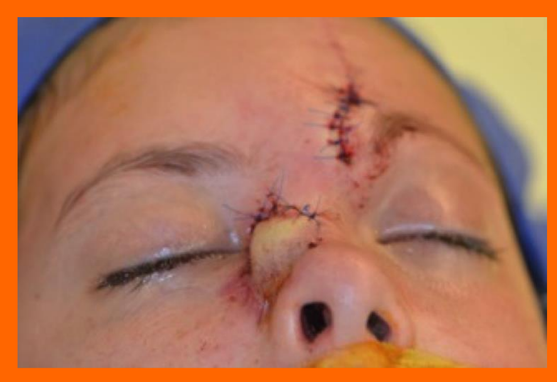


Figure 3 (Top Left) . Intraoperative photograph of resection bed from second resection at our institution.
Figure 4 (Top Right). Intraoperative photograph after third resection demonstrating the through and through defect with the nasal cavity.
Figure 5 (Bottom Right). Intraoperative photograph of patient status post second stage paramedian forehead flap reconstruction of right nasal sidewall defect.



Discussion

Spindle cell neoplasms can be diagnostic challenge given the wide range of differential diagnosis that includes benign and malignant pathology (nodular fasciitis, hemangioma, leiomyoma, lipoma, and fibromatosis, spindle cell carcinoma, spindle cell melanoma, malignant peripheral nerve sheath tumor (MPNST), synovial sarcoma, angiosarcoma, rhabdosarcoma, and other sarcomas).[1,3,4] These malignancies have been noted to appear in the larynx, oral cavity, oropharynx, nasopharynx, and maxilla.[6] Spindle cell carcinomas have been described as staining positive for epithelial markers such cytokeratin or epithelial membrane antigen as well as mesenchymal markers such as SMA, muscle specific actin, or vimentin.[1,2] The specimen from the patient's second resection stained positive for SMA, suggesting a myofibroblastic origin. Myofibroblastic tumors are also a diagnostic challenge; there are a number of benign (nodular fasciitis, fibromatosis, fibroblastomas, histioblastomas, etc.) as well as malignant (myofibrosarcoma, inflammatory myofibroblastic tumor, fibrosarcomas, etc.) lesions that demonstrate myofibroblast lineage.[7]

The management of rare pediatric malignancies is made challenging from both an oncological and psychosocial standpoint. The decision making process for our patient was made especially challenging for our multidisciplinary team given the low incidence and unclear prognosis of this type of tumor, the lack of therapeutic guidelines, and our concern for the psychosocial impact of a rare tumor requiring surgical intervention with correction of a significant facial defect in a pre-adolescent female patient. Paramedian forehead flap reconstruction is an excellent option for correcting nasal defects in children.[8] Surgical technique and timing of reconstruction are critical and are influenced by multiple factors including the psychological and social effects of an open facial wound, early or late repair, and anticipation for further nasal growth and midface maturation. Maximum nasal growth velocity for girls ranges from before 8 years of age to 12 years of age;[9] reconstruction at 6 months provided timely closure after close clinical monitoring for recurrence. The patient was provided a nasal prosthesis during this initial monitoring phase; she did not tolerate the prosthesis and we suspect that she rarely used it. This finding was consistent with reports in the literature of pediatric patients poorly tolerating facial prosthetic replacements.[8]

As a survivor of a pediatric malignancy, our patient is at risk for psychological sequelae such as depression, anxiety, and post-traumatic stress disorder.[10] The psychosocial impact of having an open wound in a cosmetically sensitive region prompted early involvement of a child psychologist. The patient and her family benefitted greatly from routine counseling and have both reacted appropriately and coped positively to the patient's condition and recovery.

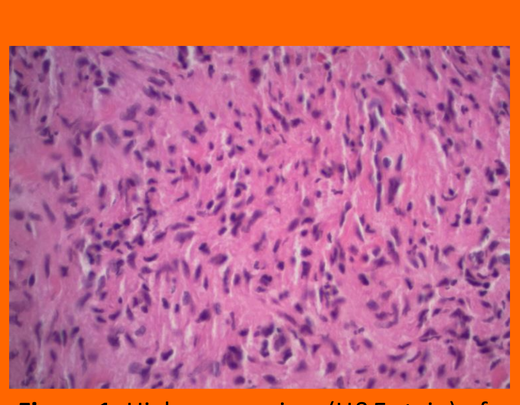


Figure 1. High power view (H&E stain) of specimen from initial resection demonstrating spindle cells.

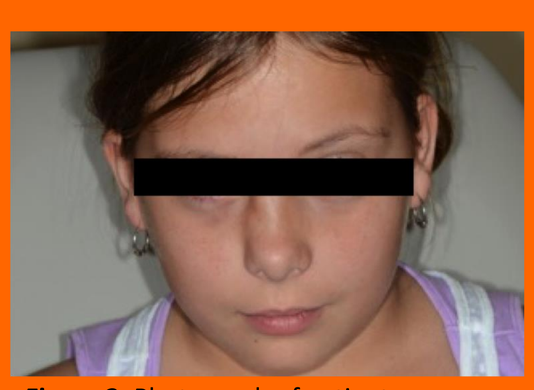


Figure 2. Photograph of patient demonstrating nasal asymmetry.

Conclusion

Spindle cell neoplasms remain a diagnostic challenge and an invasive lesion may not resemble well-known oncological diagnoses. Our patient suffered a rare, atypical, locally infiltrative spindle cell lesion on her nasal sidewall with multiple recurrences after resection. This case is unique in that our pre-adolescent patient suffered a rare malignancy causing significant cosmetic deformity that required cautious decision-making. The psychological and social impact of each management option was anticipated and discussed often. A final decision was made with the assistance of our oncology and psychology colleagues. She has not had a third recurrence of her malignancy to date. For child or adolescent patients that are suffering similar conditions with a significant defect after cancer resection, we recommend a mutli-disciplinary approach to management, frequent contact and follow up with family, and early involvement of psychology.

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