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Pleomorphic hyalinizing angiectatic tumors: Recognizing a novel source of cancer in the upper extremity

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Statement of the Problem: Soft tissue masses of the upper extremity often represent benign pathological entities, but the astute hand surgeon should use precaution when the possibility of malignancy arises. In the differential diagnosis, one needs to consider pleomorphic hyalinizing angiectatic tumors (PHAT), a novel pathology with increasing reports in the literature. The aim of this study is to report a case of PHAT in the hand and to describe diagnostic findings and management from a systematic review.

Methodology & Theoretical Orientation: We report a case of PHAT and conducted a systematic review of the literature for all published data on diagnosis and management of this novel entity. Publications from 1965 to 2016 were selected from databases such as PubMed/Medline, Cochrane Review and Google Scholar. Collected data included patient characteristics, anatomical sites of predisposition, tumor sizes, clinical signs, imaging findings, treatment options and recurrence. Results are reported as means and interquartile ranges.

Findings: In total, there have been 48 publications on PHAT, but only 8 reported it in the upper extremity including 11 patients. Tumor sizes ranged from 26 cm to 4 cm in largest diameter with clinical findings significant for pain on direct palpation in 91% of cases. Treatment strategies include surgical excision with wide margins (91%) and radiotherapy (9%), without any chemotherapy. No local recurrences have been reported for 10 years.

Conclusion & Significance: Pleomorphic hyalinizing angiectatic tumors are rare soft tissue masses that can arise in the upper extremity. Early recognition of clinical signs and surgical resection with wide margins has not resulted in recurrence so far.

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