Pleomorphic Hyalinizing Angiectatic Tumors: Recognizing a Novel Source of Cancer in the Upper Extremity

Johnny Ionut Efanov[1] MD MSc, Katherine Larose, Anne-Julie Labrecque[1] MD, Michèle Tardif[1] MD
[1]Division of Plastic Surgery, Hôpital Maisonneuve-Rosemont, Université de Montréal, QC, Canada

Case Presentation

A 60-year-old, right-handed, truck driver presented with a non-traumatic slow-growing mass on the palm of his left hand. The slow-growing mass appeared 3 years ago and worsening pain on palpation with signs of inflammation prompted his referral. He denied smoking, medication use, allergies, family history of cancer and any other associated symptom.

On hand examination, a large subcutaneous, non-mobile mass overlying the proximal wrist crease and abutting the proximal radial border of the hypothenar eminence was palpated (Figure 1). Largest superficial diameter was 4.5cm. Neurovascular signs were normal, active and passive range of motion was unaffected. Upper extremity and axillary examination revealed no lymphadenopathy.

Imaging

3-view x-ray radiography: No calcium deposits or ossifications in the tumor, underlying carpal-metacarpal bones unaffected. Soft tissue swelling on the palmar side suggested a mass effect.
Magnetic resonance imaging (MRI): Polylobulated, 4.3 X 1.4cm mass in subcutaneous soft tissue lying on superficial palmar aponeurosis.

Treatment

An elliptical skin incision over the carpal tunnel was performed. Surgical dissection of soft tissue surrounding the mass was undertaken, ensuring that wide margins of over 1cm in all directions were taken.

Pathology

Pathology report revealed a multi-lobed tumor, moderately cellular, and with several vessel ectasia with hyalinized walls. Neoplastic cells were partly fusiform with moderate atypia, and partly pleomorphic with a large multi-lobed hyperchromatic nucleus. Several cells contained hemosiderin deposits. Stroma was partially hyalinized and some calcifications were present. Mitotic activity was low. Immunohistochemistry demonstrates a positive marking for Vimentine, a focal marking with Desmine and absence of marking to CD34, S100 protein, keratin AE1-AE3 and FVIII.

Diagnosis

Final diagnosis indicated that the lesion corresponded to a pleomorphic hyalinizing angiectatic tumor (PHAT). Surgical treatment proved to be sufficient, avoiding the need for chemotherapy or radiotherapy postoperatively. No signs of recurrence noted after 18 months of follow-up.

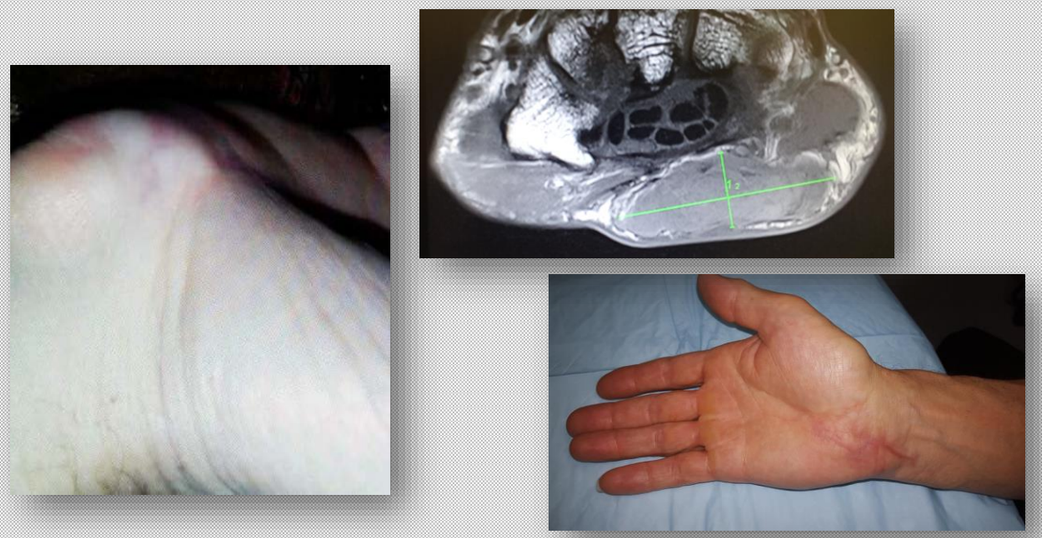

Figure 1: (*Left*) Appearance on clinical presentation. (*Upper*) MRI findings. (*Lower*) Results at 18 months without recurrence.

Table 1: Systematic review of the literature for publications reporting PHAT in the upper extremity.

Study	Year	Sex	Age	Hand Dominance	Site	Preoperative Duration	Tumor Size (cm)	Clinical Signs/Sx	Imaging Findings	Treatment	Local Recurrence
Folpe & Weiss	2004	M	32	NA	Arm	Unknown	6,0	NA	NA	NA	NA
		F	39		Hand	Unknown	Unknown			NA	NA
		M	42		Forearm	Many years	Unknown			SE, Radiotherapy	None at 5 years
		M	67		Arm	Unknown	5,8			NA	NA
Parameshwarappa & al.	2010	F	65	NA	Forearm L	13 years	5,0 x 4,0	Intermittent dull ache x 3 years	NA	SE	None at 5 years
Subhawong & al.	2012	F	87	NA	Arm R	3 years	4,8 x 4,6 5,8	Intermittent pain ameliorated with repositioning	Multiloculated cystic mass with nodular and thickened enhancing septa	SE	None at 7 months
Changchien & al.	2014	M	76	NA	Arm L	5 months	4,6 x 4,5 x 3,0	None	NA	SE	None at 2 months
Brazio & al.	2015	M	22	R	Forearm R	2 years	16,7 x 24,2 x 26,0	Pain, Arm weakness	Cystic and/or necrotic areas and linear areas representing hemorrhage or soft-tissue components	SE	NA
Michal & al.	2016	M	63	NA	Forearm	NA	4,0 x 3,5 x 2,5	NA	NA	SE	NA
Kane & al.	2016	M	35	R	Hand R	Many years	4,6 x 2,8 x 1,8	Painful with power graps and direct pressure	Consistent with a lowflow hemangioma	SE	NA
Efanov & al.	2016	M	60	R	Hand L	3 years	4,3 x 1,4	Painful with direct pressure	Polylobulated without abscess or necrotic material	SE	None at 18 months

Discussion

<100 cases of pleomorphic hyalinizing angiectatic tumors (PHAT) of soft tissue have been described to date. Most cases occur in superficial soft tissues in the lower extremities, in both pediatric and adult populations.

In our literature review (Table 1), PHAT tumors were seen in the upper extremity of adults aged between 22 and 87 y.o., predominantly in men (73%) and mostly in the arm or forearm, with only one case reported in the hand. Usually, growth of the mass occurs over extended periods of time and can reach dimensions as large as 26cm. Clinical signs are non-specific and patients usually complain of intermittent pain. Imaging studies usually demonstrate cystic masses with signs of hemorrhagic and necrotic areas.

Management options for PHAT are solely surgical for the time being, with no evidence of benefit from adjuvant chemotherapy or radiotherapy. No reports suggest a metastatic potential. Therefore, we believe that a large surgical excision with negative margins remains the best option when addressing PHAT. In the literature, recurrence rates have been shown to be as much as 30-50%, all of them following marginal excision, which reinstates the necessity for wider surgical margins. Fortunately, there have been no cases of upper extremity recurrences.

One of the difficulties with pathological diagnosis of PHAT relies in its similarities with other sarcomas. Undifferentiated sarcomas with fusiform or pleomorphic cells can resemble PHAT under microscope examination. Our patient did not express CD34 which was often seen in other reports of PHAT. It is arguable that neo adjuvant treatment could be provided in the context of uncertainty between PHAT and a sarcomatous tumor. However, the safety of performing a wide surgical excision in the upper extremity seems to be sufficient with regards to the aggressiveness of this cancerous tumor.

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