CASE REPORT

Conventional (Adult Type) Hemangiopericytoma of Thigh in Association with Arteriovenous Malformation and Epidermal Inclusion Cyst: A Rare Combination, At an Unusual Site

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Abstract:

Hemangiopericytoma was first described in 1942 by Stout and Murray as a distinctive soft tissue neoplasm of pericytic orgin characterized by 'staghorn' branching pattern. It is no longer considered a specific entity but rather a growth pattern shared by many unrelated benign and malignant neoplasms. The pathogenesis of Areteriovenous malformations is not well understood. Rarely, it has been reported with associated arteriovenous malformations. We present a rare case of combination of Hemangiopericytoma with arteriovenous malformations and epidermal inclusion cyst in the same lesion in leg. A 62 year male patient presented with a polypoidal cutaneous growth over posterior aspect of right thigh measuring 8x8x5 cm for past 25 years, firm in consistency. Radiological features revealed an areteriovenous malformation with soft tissue attenuation vascular mass. Cytological findings of the swelling were suggestive of a vascular lesion with cystic degeneration. Excision of the mass was performed and specimen was received as a large skin covered polypoidal mass measuring 14x10x5 cm. To the best of our knowledge, no prior report mentions a tumour of this size, associated with arteriovenous malformation and epidermal inclusion cyst of lower extremity.

Keywords: Arteriovenous Malformation, Epidermal inclusion cyst, Hemangiopericytoma, Vascular lesion

Introduction:

Haemangiopericytoma (HPC) is an uncommon mesenchymal neoplasm (less than 1%) predominantly involving adults often with a protracted course, first described and named by Stout and Murray in 1942 [1], however it was not until 1949, when Stout reported 25 additional cases, that the tumour received widespread recognition [2]. Over the years, it appeared that this growth pattern was a non specific one, shared by numerous, unrelated benign and malignant neoplasms and that HPC was better considered as a diagnosis of exclusion. Three categories of lesion may now be individualized within the heterogenous group of HPC-like neoplasms.

The tumours of first category are the tumours with HPC like features which include synovial sarcoma with HPC like features, second category tumours include true HPC with myoid/pericytic differentiation including HPC, infantile myofibromatosis, glomangiopericytoma/myopericytoma etc. and the third category includes the solitary fibrous tumour group comprising of solitary fibrous tumour, giant cell angiofibroma and lipomatous HPC [3].

The head and neck region is the main site of tumour location (15-30%), while involvement of lower limbs, lung, pelvis and visceral organs occurs less frequently. It is believed to arise from the pericytes, contractile spindle cells that surround the capillaries and post-capillary venules [1-2, 4, 5].

The tumour typically comprises of uniform elongated cells surrounding a rich, branching network of thin-walled vessels of various sizes and shapes [6]. It occurs commonly in fourth to fifth decade of life and shows no sex predilection. HPCs may be benign or malignant, have been identified in all age groups, demonstrate equal gender distribution, and have a high propensity for metastasis [4, 5]. Spread is not a common feature of the tumour and reports of spread occur frequently after incomplete surgical resection [5]. Grossly, presents as well circumscribed nodular mass. Microscopy shows unencapsulated tumour tissue, well circumscribed, comprised of cytologically uniform small, basophilic, ovoid to spindle shaped cells. These are arranged in a pattern less fashion around thin walled blood vessels, which are in a staghorn configuration [7].

Arteriovenous Malformation (AVM) is an abnormal connection between arteries and veins, bypassing the capillary system. The pathogenesis of AVMs is not well understood. Intracranial AVM is the most common, followed by extracranial head and neck, extremity, truncal, and visceral sites. AVMs are usually noted at birth but are frequently misdiagnosed. Usually, they are mistaken for a hemangioma. Fast-flow typically becomes evident during childhood. The mass occasionally shows rapid enlargement following trauma or during puberty. Later consequences of arteriovenous shunting include ischemic signs and symptoms and indolent ulceration. Clinical diagnosis is confirmed by computer tomography, ultrasonography and colour doppler examination. Grossly, presents as tangled mass of dilated tortuous vessels. Histopathological features show thick and thin blood vessels corresponding to arteries and veins [7, 8]. Epidermal inclusions cysts present as discrete nodules with the wall composed of mature squamous epithelium, which may be formed from areas of previous trauma which may cause the epidermis to sequester in the dermis allowing for slow growth of the lesion through sloughing of dead cells centrally [9]. The objective of the study is to present a rare case with combination of hemangiopericytoma with arteriovenous malformations and epidermal inclusion cyst in the same lesion in leg. Thus it is unique, because of coexistence of three entities in the same lesion.

Case Report:

A 62 year male patient presented with a polypoidal cutaneous growth over posterior aspect of right thigh measuring 8x8x5 cm for past 25 years increasing in size, firm in consistency. Contrast enhanced CT of right thigh revealed an AVM with soft tissue attenuation vascular mass (Fig. 1). Cytological findings of the swelling were suggestive of a vascular lesion with cystic degeneration. Surgical excision of the mass was performed.

Pathological findings:

Specimen was received as a large skin covered polypoidal mass measuring 14x10x5 cm, external surface showed a focal ulcerated area close to the pedicle of mass (Fig. 2). Cut surface showed partially solid and partially cystic areas, solid areas were soft, tan brown. Cystic areas were filled with blood clots and comprised of thrombosed areas (Fig.3) This cyst was located in the superior-medial location measuring 3.5x3 cm. Sections stained in haematoxylin and eosin, from the solid areas showed staghorn endothelium lined vascular channels surrounded by spindle to oval cells with elongated nuclei, vesicular chromatin, mild anisonucleosis and tapering eosinophilic cytoplasm arranged in sheets**.** Reticulin showed positivity for reticular fibres encasing the tumour

cells in perivascular pattern (Fig.4a, b, c). Section from the stalk showed thin and thick walled arteries and thick arteriolised venous channels, compatible with AVM (Fig.5). The cyst was lined by stratified squamous epithelium, compatible with epidermal inclusion cyst (Fig.6a, b)

IHC Study (Clone Retrieval):

Tumour cells were diffusely immunopositive for Vimentin, CD99 (Focal) (Fig.7a,b) and negative for CD31,Bcl2,CD34,S-100, CK, EMA and SMA (Fig.8a,b).

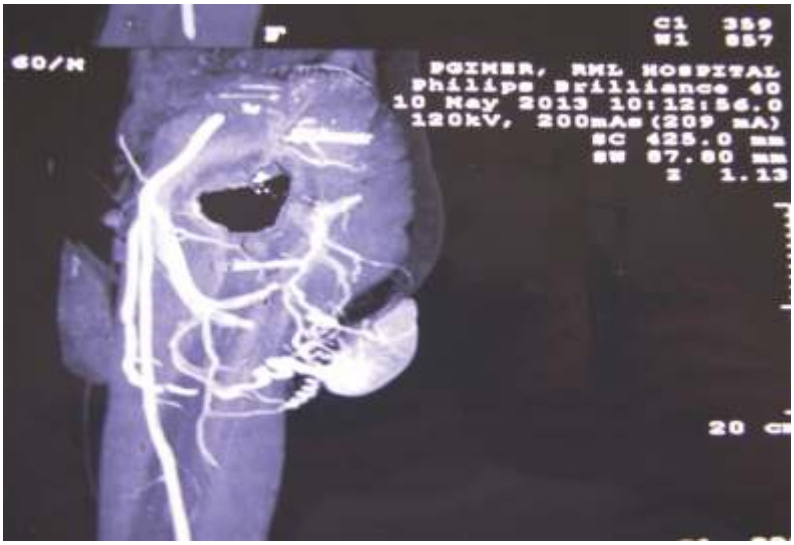

Fig. 1: Contrast Enhanced CT of Right Thigh Revealed an Areterio-Venous Malformation with Soft Tissue Attenuation Vascular Mass

Fig. 2: Skin Covered Polypoidal Mass, Showing Focal Ulceration near the Pedicle

Fig. 3: Cut section shows Cystic and Solid Areas, Cystic Areas Appearing Thrombosed

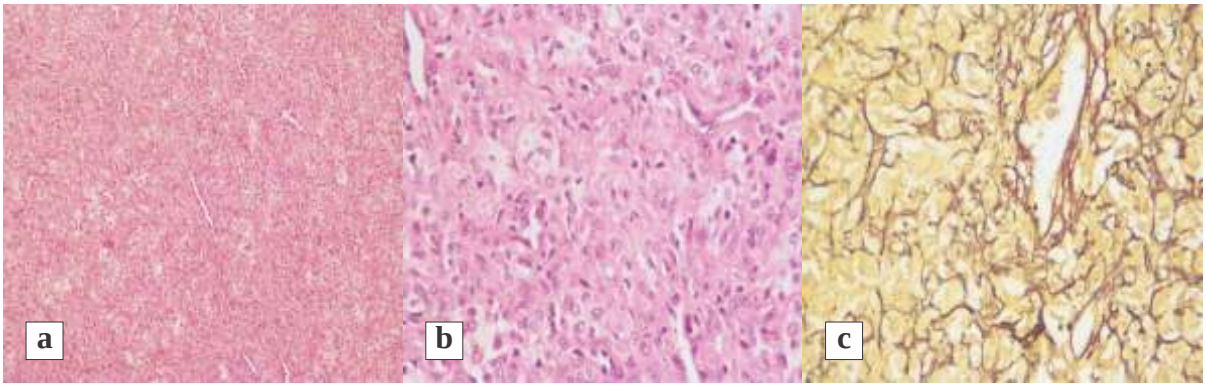

Fig. 4a: Tumour Tissue showing Staghorn Endothelium Lined Vascular Channels Arranged in Sheets (100X, H and E)
Fig. 4b: Few Staghorn Shaped Vessels Surrounded by Spindle to Oval Cells with Elongated Nuclei, Vesicular Chromatin, Mild Anisonucleosis and Tapering Eosinophilic Cytoplasm (400X, H and E)
Fig. 4c: Reticulin Stain showing Reticular Fibres Encasing the Tumour Tissue (400X)

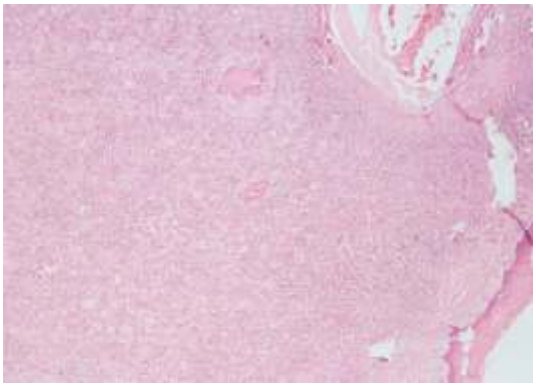

Fig. 5: Ectatic Thickening of Venous Wall (Arteriolization of vein), Thickening of Vessels and Proliferation of Thick and Thin Walled Arterioles and Venules and Part of Skin Surface (100x, H and E)

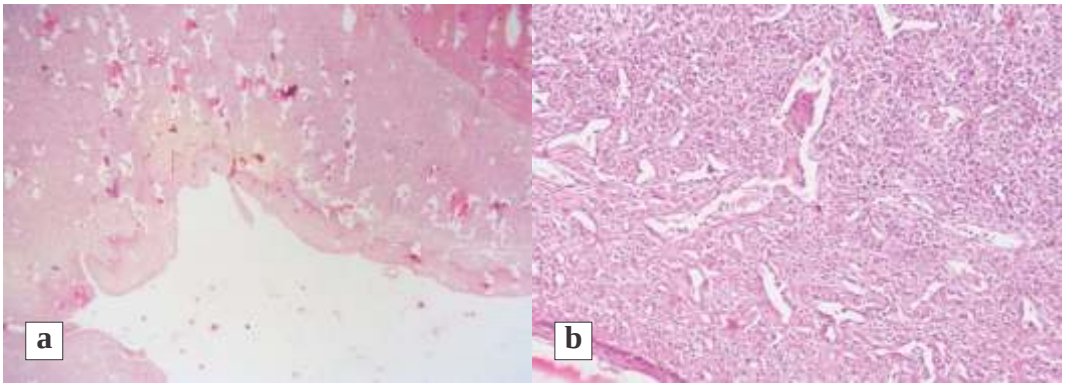

Fig.6a: Cyst Wall Lined by Keratinized Stratified Squamous Epithelium along with Thick and Thin Walled Vessels Along with Area of Venous Thrombosis.
Fig. 6b: Cyst Wall with Tumour Tissue (100X, H and E)

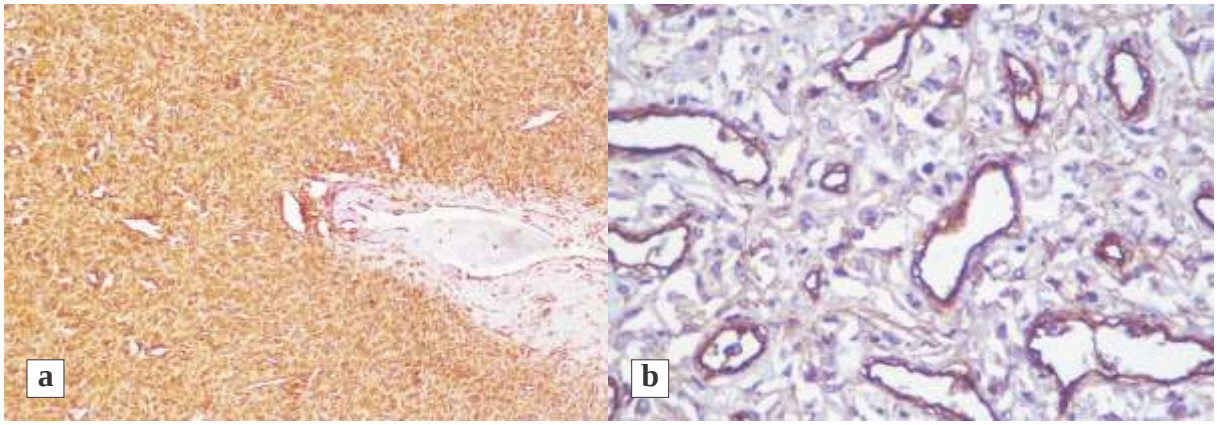

Fig. 7a, b: Vimentin-positive and CD99- Focal and Weak positive (100X)

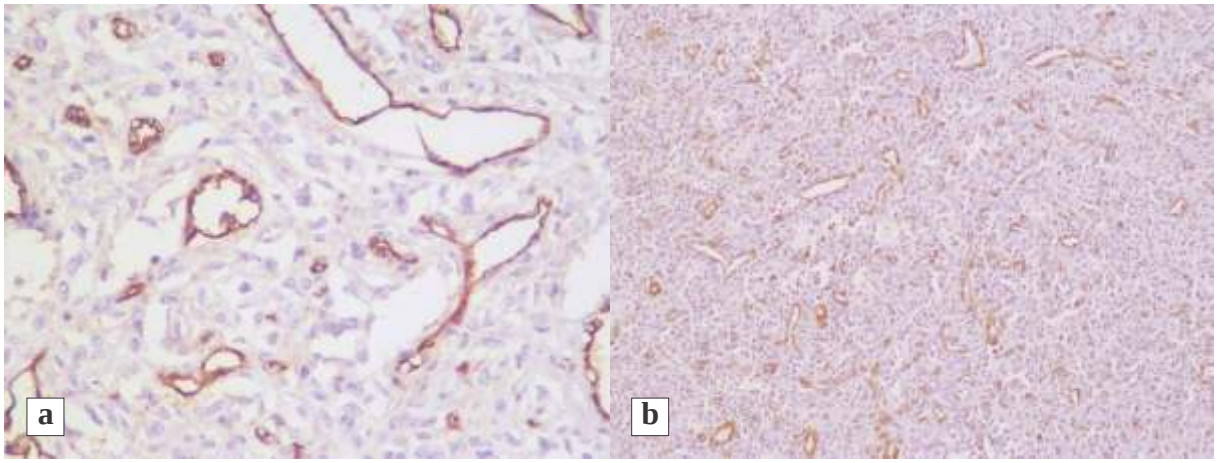

Fig. 8a, b: CD34-Negative (400X), SMA- Negative (100X)

Discussion:

HPC accounts for less than 2% of soft tissue sarcomas [10, 11]. It may occur at any age but is most common in the fifth and sixth decades [4, 5]. In the series of 106 patients reported by Enzinger and Smith, 37(35%) cases affected the lower extremity. It was first described in 1942 by Stuart and Murray as distinctive soft tissue neoplasm, presumably of pericytic origin, exhibiting a characteristic well developed 'staghorn' branching vascular pattern.

AVM may either be congenital or acquired. Most physicians prefer the term Arteriovenous Fistula (AVF) for acquired lesions and AVM for congenital lesions [12]. Acquired AVFs are almost exclusively the result of a penetrating trauma caused by a knife, bullet, or other missile with simultaneous injury to an adjacent artery and vein [13]. Because traumatic iatrogenic vascular injuries occur most often in the extremities, the arms and legs are the main sites for AVF. The penetrative injuries lead to abnormal endothelial lined communication with artery and vein with no interposed capillary bed [14].

In study by Kalani *et al*. (2011) have described an intracranial HPC found in association with multiple arteriovenous malformations and dural arteriovenous fistulae. They postulated the possibility of the hemodynamic constraints of the occluded superior sagittal sinus and the aberrant physiology of the pericyte which may have contributed to the vascular malformations [15]. In study by Kurisu *et al.* (2014) have presented a case of dural AVF associated with hemangiopericytoma that was located entirely inside the dural sinus [16]. Epidermal inclusion cysts can be found wherever there is squamous epithelium, presumably from the sites of trauma including surgery. Trauma is thought to result in sequester of the epithelium in the subepithelial tissue with subsequent slow growth through central accumulation of sloughed dead cells [8]. In case report by Koukourakis *et al.* (2015) HPC showed positivity for Vimentin, Bcl-2, CD-99 and Negative for SMA of the tumour cells [17]. However, in study by Kitahata *et al.* (2010) reported CD99 and bcl-2 negative in their case report of hemangiopericytoma of sacrococcygeal space [18]. Currently, there is a consensus that solitary fibrous tumour is a distinctive neoplasm that is barely distinguishable from HPC on light and electronic microscopic examinations. Ultra structurally, both lesions display varying features of pericytic, fibroblastic and/or myofibroblastic differentiation. Immunohistochemically, SFTs, especially the fibrous form, commonly express CD34 (80-90% of cases) and CD99 (70%), Bcl-2 (30%), Epithelial Membrane Antigen (EMA) and Smooth Muscle Actin (SMA) (20%) may occasionally be expressed. They are usually negative for S-100 protein, desmin and cytokeratins. Cellular (conventional) forms of SFT tend to be less frequently positive for CD34 [3]. Although recently case reports of isolated HPC have still been published, this case was diagnosed as conventional HPC based on the clinical, histopathological and immunohistochemistry features.

Conclusion:

HPC is a rare entity and its association with AVM occurring at uncommon site, involving the right thigh makes it an interesting case to present. Clinico-radiological features thus helped us aid to clinch the preliminary diagnosis, but cytological features and final histopathological characteristics aided with immunohistochemistry helped to confirm the diagnosis. To the best of our knowledge there is no case report showing association of HPC with epidermal inclusion cyst. Its unusual clinical presentation at an uncommon site made the case rare and very interesting and trauma was possibly the common etiological factor for development for all the three entities in our case report. Hence, we are presenting this case.

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