Hemangiopericytoma of the Hand

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Introduction

Hemangiopericytoma is a rare vascular tumour originating from the uncontrolled proliferation of pericytes and are typically being low grade benign sarcomas composed of proliferating capillary pericytes. They are mesenchymal tumours that account for 3 to 5 % of all soft tissue sarcomas and 1 % of all vascular tumours [1, 2]. These tumours have a wide anatomical distribution but tend to develop in subcutaneous tissue and skeletal muscle. 34.9 % of all hemangiopericytomas affects the lower limb, where as 15 % affect head and neck region [2]. Due to its vascular origin, it can be found anywhere in the body [3]. Hemangiopericytoma of the upper extremity is rare [1, 4]. We report a case of hemangiopericytoma of the dorsum of hand in a 50 year old male patient because of rarity and rare site of occurrence.

Case Report

A 50 year- old- male patient, known hypertensive and diabetic, presented to surgical outpatient department with a swelling on the dorsum of his right hand for 2 months. It was insidious in onset and gradually progressed to attain the present size. There was no pain or skin change associated with the swelling. The patient consulted a local doctor, who suspected an abscess. Since the patient's blood sugar

remained high he was referred to our hospital for further management.

Local examination revealed a non- tender, warm, lobulated swelling of 6×3 cm over the dorsum of ring and little finger of right hand. Skin overlying the swelling was normal. The swelling had restricted mobility and was compressible. (Fig. 1) Systemic examination did not reveal any abnormality.

His routine blood investigations were within normal limits. Sonography of soft tissue of right hand showed lobulated soft tissue mass measuring 4.3×3 cm predominantly hypo echoic showing intense vascularity with prominent surrounding arterial feeders arising from the extensor aspect of the distal hand, extending till the level of the first metacarpo phalangeal joint overlying the fourth digit. These features were suggestive of probably haemangioma. MRI revealed a large lobulated enhancing mass measuring approximately 8×4×3 cms which showed heterogenous hyper intense signal on T2 4 STIR sequences with multiple flow voids within was seen arising from the dorsal aspect of the right hand in relation to the extensor aspect of fourth and fifth digits. Proximally, the mass was extending from the level of the distal carpal row insinuating along the space between the fourth and fifth digits. Distally, the mass was extending up to the middle phalanx of the fourth digit. The lesion was encasing the extensor digitorum minimi and extensor digitorum tendons and the accompanying digital neurovascular bundle. The mass was involving the fourth dorsal interosseous muscle. No obvious extension in to the flexor compartment was noted. The underlying bones showed no altered marrow signal intensity or cortical destruction. MRA depicted the hypertrophied radial and ulnar arteries and prominent palmar arch with tortuous arterial feeders arising from the common digital palmar

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Fig. 1 Photograph of the patient showing the tumour on the dorsum of left hand

arteries (4th and 5th). Based on these findings a vascular soft tissue tumour (haemangioma) was suspected.

Wide local excision of the tumour with ligation of all feeding vessels and primary closure of the wound was done under general anaesthesia. (Fig. 2a and b). Histopathological evaluation of the resected specimen showed a thin walled endothelium lined vascular spaces around which tightly packed tumour cells. The tumour cells were uniform with eosinophilic cytoplasm, round to oval nuclei, high n: c ratio, vascular nuclei and occasional prominent nucleoli. Occasional mitotic figures were seen. (Fig. 3). The tumour specimen was positive for CD34 and vimentin and negative for Bcl-2, CD99, c-kit, factor VIII, desmin, alpha-smooth muscle actin, S-100 protein, epithelial membrane antigen and keratin upon immunohistochemical analysis. On reticulin special stain, the individual cells were surrounded by reticulin (Fig. 4). These features confirmed the diagnosis of hemangiopericytoma. Patient received 1 month of adjuvant radiotherapy following surgery as per oncologist's advice. Patient is asymptomatic on 12 month's follow up without any local recurrence or distant metastasis.

Discussion

Hemangiopericytoma is a rare vascular tumour originating from the uncontrolled proliferation of pericytes.

Fig. 3 Photomicrograph showing the tumour

Stout and Murray [1] first described it in 1942. Histologically, its main feature is the presence of clusters of small ovoid cells around capillaries and post capillary venulae, which are called pericytes. These cells are supposed to derive from perivascular pluripotent mesenchymal cells. About 1 % of all vascular tumours are hemangiopericytomas. Such tumours have a wide anatomical distribution but tend to develop in subcutaneous tissue and skeletal muscle. 34.9 % of all hemangiopericytomas affect the lower limbs, whereas 15 % affect the head and neck region [2]. Hemangiopericytoma of upper extremity is rare [1, 2]. There are 83 cases reported in English literature, of these only 14 are documented as malignant [4]. Hemangiopericytoma accounts for less than 2 % of soft tissue sarcomas. Hemangiopericytoma is primarily a tumour of adults and is rare in infants and children. There is a long natural history. It may occur at any age but it is more common in the 5th and 6th decade. It occurs equally in both sexes. It occurs most commonly in the soft tissues of the upper and lower extremities, pelvis, and retroperitoneal space and seldom in visceral organs [5]. Due to its vascular origin, it can be found anywhere in the body [2]. The etiopathogenesis of hemangiopericytomas is not yet defined. Factors such as previous trauma or a positive anamnesis after treatment with corticosteroids have been considered as possible causes of hemangiopericytomas.

Fig. 2 **a** Intra-operative Photograph showing resection of tumour. **b** Photograph of resected specimen

Fig. 4 Photomicrograph of Reticulin stain (Brown borders in the slides are reticulin stain) in which individual cells are surrounded by reticulin stain

In the series of 106 patient reported by Enzinger and Smith [6]; 35 % cases affected the lower extremity and 25 % were in pelvis or retroperitoium. Of the other 43 cases in that series, 17 involved the head and neck region, 15 were located in the trunk and 11 were in the upper extremity. Hemangiopericytoma generally present as a painless mass, but pressure on adjacent viscera may cause symptoms [5]. In our case the patient presented as painless swelling. The reported frequency of calcification in hemangiopericytoma varies from one of 106 in the series of Enzinger and smith to 5 of the patients described by Alpert et al [5]. In our case there was no calcification. Bone erosion or cortical destruction was not seen in our patient. Diagnosis of hemangiopericytoma is based on the histological and immunohistochemistry aspect. Microscopically the tumour consists of numerous vascular spaces of variable size and shape separated by aggregates of tightly packed oval to spindle-shaped cells with bland nuclei. Mitotic figures and nuclear atypia are variable in number and degree [4]. Pathologically to differentiate between of a hemangiopericytoma and other mesenchymal tumours such as solitary fibrous tumours can be difficult. Bcl-2 and CD99 immunohistochemistry is used to differentiate between these two tumours. Bcl-2 and CD99 markers are positive in solitary fibrous tumour [4, 6]. Reticulin (special stain) is used to differentiate hemangiopericytoma from hemangioendothelioma. In hemangiopericytoma, individual cells are surrounded by reticulin whereas in hemangioendothelioma, group of cells are surrounded by reticulin [4, 6].

Hemangiopericytoma is a benign tumour, although it has a malignant potential and a high frequency of relapse. Surgery should be as radical as possible to avoid incomplete tumour resection, the high frequency of relapse and a higher risk of malignancy. The present treatment includes complete en bloc excision of the mass with its capsule to avoid the risk of relapse [2]. Although unusual, hemangiopericytoma can be considered

benign or malignant depending on histological features. In some cases, malignant hemangiopericytoma can behave as high-grade sarcomas. When it is deemed malignant, radical surgical excision is the treatment of choice. Adjuvant chemotherapy and radiation therapy offer limited success in patients with this malignancy [7].

Hemangiopericytomas are reported to occur at many other sites in the body including intraosseous, nasal cavity and sinuses (Sino nasal) meninges. Retroperitoneal and meningeal tumours had a higher local recurrence rates when compared to extremity tumours which are significantly better. Histological features of aggressive biological behaviour are presence of necrosis, mitoses, vascular invasion and pleomorphism. In our case high n: c ratio and occasional mitotic figures were seen. High grade lesions are the one which usually metastasize. The most common site of metastasis being the lungs and the less common sites of metastasis are bones, lymph nodes and liver [8]. Metastatic disease is usually the cause of death in these patients.

In both adults and children, curative surgery is the most important predictor of survival. Complete surgical resection remains the mainstay of treatment for hemangiopericytoma [2, 4, 9]. The place of chemotherapy in the treatment of this tumour is not well established [2–6]. Treatment is primarily surgical with adjuvant radiotherapy for tumours in location like retroperitoneal and meningeal tumours which have a higher recurrence rates [8]. Because of unpredictable biological behaviour of adult-type, supplementary treatment may be considered individually. In unresectable adult-type adjuvant chemo- and radiotherapy should be administered [2, 4, 7, 8]. Chemotherapy can help in achieving partial or complete responses in metastatic hemangiopericytomas. The drugs used are actinomycin-D, adriamycin, cyclophosphamide, methotrexate and vincristin [8]. But the prognosis is poor despite supplementary treatment. High chemo responsiveness of the infantile-type provides favourable outcome even in cases of unresectable, life-threatening tumours. Complete remission after chemotherapy alone is not infrequent [7].

Conclusion

Hemangiopericytoma of upper extremity is rare. Because of the unpredictable behaviour of this tumour, extended follow up are strongly recommended. Complete surgical excision remains the mainstay of treatment in both infantile- and adult-type. Because of unpredictable biological behaviour of adult-type, supplementary treatment may be considered individually. In unresectable adult-type adjuvant chemo- and radiotherapy should be administered.

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