Case Report

Spindle cell hemangioma- A rare case entity

Jenita Christiana[1*], Meharaj Banu O A[2], Arunkumar N[3], Uma R[4], Prathipa K[5]

[1]Associate Professor, [2,3,4,5]Assistant Professor, Department of Pathology, Government Thiruvarur Medical college, Vilamal, Thiruvarur – 610004 **Email:** bansarind@gmail.com

Abstract

Spindle cell hemangioma is a rare vascular tumor common in extremities. Earlier it was described as spindle cell hemangioendothelioma but later because of its benign indolent course, it was renamed as spindle cell hemangioma. We report two cases of SCH who are 35 yrs and 55 yrs male, in preauricular region and shoulder region respectively. Though this tumor is common in distal extremities, we report two cases, one in the shoulder region and other over the face in right preauricular region. The histopathological report of both shows cavernous spaces and spindle cell areas. These cases are reported here due to its rarity of site.

Keywords: Spindle cell Hemangioma, Spindle cell hemangioendothelioma, Cavernous space, Spindle cells, Vascular tumor.

[*]**Address for Correspondence:**
Dr. Jenita Christiana, Associate Professor, Department of Pathology, Government Thiruvarur Medical College, Vilamal, Thiruvarur – 610004, Tamil Nadu, INDIA.
Email: bansarind@gmail.com
Received Date: 03/10/2015 Revised Date: 27/10/2015 Accepted Date: 31/11/2015

Access this article online

Quick Response Code:

Website: www.statperson.com

DOI: 12 December 2015

INTRODUCTION

Spindle cell hemangioendothelioma was first described by Weiss and Enzinger in 1986[1,2,3]. It was regarded as a vascular tumor of low grade malignancy[4,5,6]. Results of larger studies with various investigations and longer follow up indicated the clinical behavior of these tumors. Hence currently this is viewed as benign neoplasm and renamed as Spindle cell hemangioma[3,6] by WHO in 1996[5,8]. Spindle cell hemangioma have a wide age distribution[2,4,7] although it often presents in children or young adults[1,2,7]. Male and female are affected equally[2,6]. They usually present as firm, red -blue subcutaneous and dermal nodules that may be solitary or multifocal[3,6], most often on the distal aspect of the extremities [1,2,4,6,7]. The other sites of occurrence includes chest wall, genital area, head and neck[5,8], oral cavity[3,5,9,10] and nasal cavity[8]. It has also been found in the viscera, spinal cord and cervix[3]. We hereby report two cases of spindle cell hemangioma, one in the preauricular region over the face and the other in the shoulder.

CASE REPORT

Case 1: 35 yrs old male patient presented with chief complaint of swelling near the right ear since 7 months. Initially swelling was presented as small nodule and gradually increased to present size, with the history of pain for 1 month. Fine needle aspiration cytology was done and it showed only blood and its components. On examination, swelling was 3 x 2 cm in size, firm in consistency, tender and slightly mobile. Excision biopsy was done and sent to our department. We received single globular soft tissue mass measuring 4 x 3 x 2 cm with congested external surface. C/S shows predominant brownish with central grey white area.

Case 2: 55 yrs male presented with the complaint of gradually enlarging tumor over the left shoulder region since 1 year duration with the history of pain for 2 months. Pain aggravated on lifting the left upper limb. Fine needle aspiration cytology was not done for this patient. On examination, swelling was 5 x 4 cm in size with firm consistency, tenderness and slight mobility. The surface appears to be smooth. X-ray showed a soft tissue swelling with no bony abnormality. Excision biopsy was done. We received single skin covered soft tissue mass measuring 6 x 5 x 3 cm. C/S shows predominant cystic areas filled with hemorrhage and focal solid areas. Histopathological examination of both shows numerous varying sized cavernous spaces lined by attenuated

How to site this article: Jenita Christiana, Meharaj Banu O A, Arunkumar N, Uma R, Prathipa K. Spindle cell hemangioma- A rare case entity. *International Journal of Recent Trends in Science and Technology.* December 2015; 17(2): 97-100. http://www.statperson.com (accessed 12 December 2015).

endothelial cells showing organising thrombi and frequent phleboliths intervened by solid areas composed of spindle cells, epithelioid cells with intracytoplasmic vacuoles, slit like blood vessels, hemosiderin laden macrophages and a focal collection of perivascular lymphoid aggregates. Both cases were reported as Spindle Cell Hemangioma.

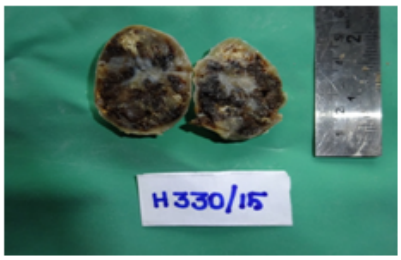

Figure 1

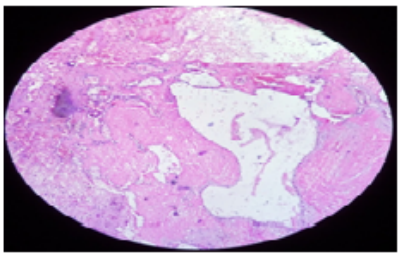

Figure 2

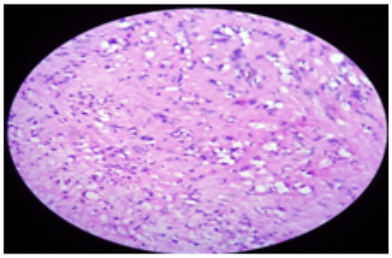

Figure 3

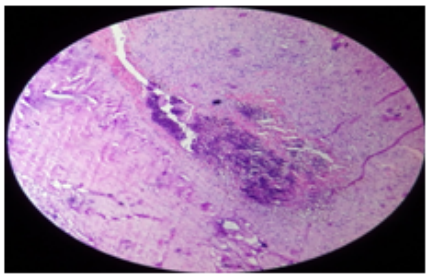

Figure 4

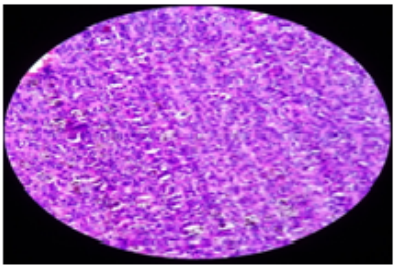

Figure 5

Legend

Figure 1: Grossly shows predominant grey brown with central grey white area
Figure 2: Section shows varying size cavernous spaces lined by attenuated endothelial cells
Figure 3: Section shows epithelioid cells with intracytoplasmic vacuoles
Figure 4: Section shows blood vessel with calcification and adjacent epithelioid cells
Figure 5: Section shows solid area composed of spindle shaped cells and epithelioid cells

DISCUSSION

Spindle cell hemangioma is a rare benign vascular tumor that can occur as solitary nodule or gives rise to multiple lesions in the same anatomic region specially when it is an intravascular growth[5]. Approximately 10% of cases has been reported to occur in association with various developmental anomalies and syndromes[5,6]. Several cases have been described with Maffucci's syndrome[6,7,8] that too multifocal. The other associations include Klippel Trenaunay syndrome, early onset varicosities and congenital lymphedema[3,5,6,7,8]. Spindle cell hemangioma may develop at any age but it has the propensity for children and young adults[1,2,4,7] without any sex predilection[2,6]. In our case the incidence was in 35 and 55 years male patients. Commonly the tumor affects the distal limbs though its occurrence has been reported in other sites such as chest wall, genital area, head and neck,[5,8] oral cavity[3,5,9,10] and nasal cavity[8] including viscera such as pancreas and spleen as well as spinal cord[3]. Unusual osseous involvement has been rarely documented by Sylvia *et al*[3] and Doo *et al*[1]. As per the reviews of Vijayalakshmi *et al* only 10 cases of Spindle cell hemangioma have been reported in the head and neck region[8]. We too reported one case in the preauricular region over the face and other in the shoulder region – proximal part of upper limb in contrast with the literatures that emphasize distal predilection. Scott and Rosai have reported one case of Spindle cell hemangiomatosis involving the whole of upper extremity[3]. There has been a considerable debate regarding the pathogenesis of this tumor and it is yet to be fully elucidated. Fletcher *et al* advocate that the lesion arises in the area of abnormal vasculature[3]. This idea is supported by Vijayalakshmi *et al* stating that variation in blood flow gives rise to alternating areas of vascular expression and collapse[8]. Imayama *et al* propose that injury incites these lesions causing reactive vascular proliferation[11] from a cycle of recanalization after thrombus and most of the cells appear simply to be fibroblasts with features of pericytes[1,3]. Eltarky *et al* also suggested SCH to be a non-neoplastic lesion based upon the facts of repeated presence of organized intravascular malformations[1]. However with its clinical behavior and histological findings it is unlikely to attribute this to be a reactive vascular change. Three classic histologic features are found in variable proportions in any given case. They are widely dilated

vascular spaces, solid spindle cell areas and plump round or polygonal endothelial cells with clear often vacuolated cytoplasm. The cavernous vascular spaces often contain thrombi, phleboliths and endothelial lined fibrous papillae. The solid kaposiform areas are composed of spindle cells in haphazard or short fascicular pattern surrounding rounded or compressed slit like vascular spaces. There are different views on the nature of spindle cells suggesting it to represent primitive endothelial cells by Weiss and Enzinger in 1986, pericytes by Scott and Rosla in 1988 or fibroblasts by Imayata in 1992[1]. Focal presence of plump or round epithelioid cells either in clusters or lining vascular channels with sometimes vacuolar changes depicting intracytoplasmic lumen formation by single cell are observed. The differential diagnosis of SCH include other types of haemangiomas [cavernous, histiocytoid, disseminated lobular capillary haemagiomas], dermatofibroma, Kaposi's sarcoma, kaposiform haemangioendothelioma and intravascular papillary endothelial hyperplasis[2,3,5]. The combination of cavernous spaces with organized thrombi, plump endothelial cells and multitude of spindle cells histologically differentiates it from other diagnosis. In the observations of Immunohistochemical analysis conducted by various studies, there was a positive staining for CD34, Factor VIII related antigen, Vimentin and lectin binding Ulex europaeus agglutinin I by endothelial cells lining vascular channel and vacuolated or epithelioid cells[4,6,8]. Spindle cells in solid areas were negative for endothelial markers[4,6,8] except for vimentin and showed divergent positive immunoreactions of HHF35, α smooth muscle actin, desmin and collagen IV[6,8]. An extracellular matrix molecule Decorin, member of leucine rich proteoglycan gene family is expressed in most collagen rich mesenchymal tissues and regulates endothelial cell matrix interactions during angiogenesis [upregulated in angiogenesis of inflammatory processes]. Recent publications demonstrate different expression patterns on benign and malignant vascular tumors. There is growing evidence to indicate that Decorin plays a diagnostic significance such that malignant vascular neoplasms like Kaposi sarcoma, angiosarcoma completely lack immunorectivity for Decorin whereas benign vascular tumors like hemangiomas readily express Decorin in detectable amounts[4,5]. Kyle.C.Kurek *et al* in their mutation analysis have detected mutation in exon 4 of the isocitrate dehydrogenase genes IDH1 or IDH2 in 71% of spindle cell haemagiomas and concluded it to be highly specific for this entity. In addition to this another highly specific genetic alteration IDH1 R132C mutations has been identified in lesional tissues and it has been recommended as a diagnostic aid for SCH[7].

CONCLUSION

SCH is a rare benign vascular tumor, previously thought to be a low grade malignancy. Exceptionally few cases of SCH with aggressive clinical behavior such as local metastasis has been reported by Silva *et al* and Lai *et al* [1]but they have not progressed to widespread dissemination. Moreover there has been no reported patient mortality or metastasis from SCH in the literature [3]. This resulted in failure to recognize this entity as low grade malignant or borderline lesions since metastasis is the diagnostic criteria required for malignancy. Approximately 60% of SCH have tendency to recur, [6,8] most likely due to the inherent multifocality of these lesions[6]. The standard treatment is wide surgical excision as limited excision almost certainly reoccur. Keeping in view the morbidity and slow growth rate of tumor, conservative excision on a case by case basis can be decided. In case of aggressive or difficult to surgically excise lesions, radiotherapy low dose interferon α-2b, Intralesional and intraarterial administration of recombinant Interleukin-2 has been successful. In light of available literatures it is important to counsel the patient with SCH on the high risk of re-occurrence and provide appropriate follow up.

REFERENCES

1. Doo Hyun chung MD, Jooseobkeum MD, Geon Kook leeMD, Chong Jae kin MD, Seomg Hoe Park Md, " Spindle cell haemangiendothelioma – A case report " journal of Korean medical science Vol 10, No 3, 211-215, June 1995
2. KapilDev Das KaushikGhosh, chinmoykar, Ramesh Chandra Gharami, sir chalerabortAmbarish Bhattacharyya, " spindle cell Hemangioendothelioma: Rare clinical entity " Indian journal of Dermatology.
3. Sylvia S.Gray, Mahmoud A.Eltroky, Roy F.Riascos, Richard D.Montilla, "spindle cell hemangioma reoccurrence in the hand case report: HAND (2012)7: 194-199
4. DaikiRokunobe, Hitoshi, Takeda, Takabidekakeno, Takayuki, Aizu, Eijiro, Akasaka, YasushiMatsuzaki, Hajimanakano, Daisuke sawamura "Spindle cell hemangima and Decorin Expression" Journal of cosmetics Dermatological sciences and applications 2012,2,8-10
5. SunandaChavva, M.Himapriya, KomaliGarlapati, G.Siva Prasad reddy, AS halite Ganepalli, "Rare case of spindle cell Hemangioma " Journal of clinical and diagnostic research, 2015, june, vol 9(6), 19-21
6. Michael E.Ming, MD " Multiple violaceous Nodules in a 46 –yrs old man – Diagnosis: Spindle cell Hemangiomas" Arch Dermato2006; Vol 142(5): 641-646
7. Kyle C.Kurek, Twinkalc.Pansuriya, Maayke A.J.H Van Ruler, BrendyVandenAkker, Valeria L.Luks, sofie L.J, Verbeke, Harry p.Kozakewich, Rafsciot Dina Lev Alexander J.Lazar, Christopher D.M Fletcher, Judith

V.M G Bovee " R132c IDH 1 mutations are found spindle cell Hemangiomas and not in other vascular tumors or malformations. The American Journal pathology Vol: 182, No ;5 May 2013:1494 -1500

8. Vijayalakshmi surangi, anil S harugopHema B Bannur, Ganga S pilli, RS mudhol "spindle cell hemangioma of the nasal cavity: A rare tumor with unusual presentation ; ClinRhin an Int J 2013:6(3):149-151
9. Sheehan M, shelia O, Roumpf: spindle cell hemangiomas ; Report of case presenting in the oral cavity J.Cutanpatho 2007 34(10): 797- 800
10. Tosios K. Koutlas IG, Kapranos N "spindle cell hemangiomna of oral cavity.A case report: J oral pathol Med. 1995;24: 379- 82
11. Imayama s, Murakamai Y, Hashimoto H, etal " sindle cell Hemangioendothelioma exhibits vascular proliferation rather that of angiosarcoma: AM J path 1992;97: 279-87

Source of Support: None Declared
Conflict of Interest: None Declared