CORPUS PUBLISHERS

Advance Research Journal of Cancer (ARJC)

Research Article

Malignant Hemangiopericytoma in Thigh: A Case Report and Literature Review

Amani Saleh Hadi[1]*, Rafaat Al Areqee[2] and SadikSaif Mohammed Aklan[3]

[1]*Specalisit of clinical oncology and nuclear medicine, National oncology center, Yemen*
[2]*Histopatholgy department, Alamal cancer center, Yemen*
[3]*Alamal cancer center, Taiz, Yemen*

Volume 1 Issue 1, 2020

Article Information
Received date: April 01, 2020
Published date: May 07, 2020

***Corresponding author**
Amani Saleh Hadi Saeed, Specalisit of clinical oncology and nuclear medicine, head of women care unit-National oncology center, Aden, Yemen

Keyword
Hemangiopericytoma; Sarcoma; Tumour; Chemotherapy

Abstract

Hemangiopericytoma (HPC) is an uncommon soft tissue sarcoma of vascular origin. It can develop anywhere there are capillaries; hence, it causes many unique problems of diagnosis and therapy. It occurs more frequently in the extremities than elsewhere in the body although it can arise in any organ. The tumour may develop over period of months and even years and in patients of all ages equally in men and women. The most critical aspect of treatment is an accurate histological diagnosis separating it from other angiosarcomas and benign tumours of blood vessels such as the solitary glomus tumour. Wide surgical excision is mainstay of treatment. However, adjuvant radiotherapy and chemotherapy are desirable because the malignant nature of this tumour is frequently unpredictable. Adjuvant therapy recommended for metastases, recurrence and incomplete resection long term fellow up is essential in all cases as recurrence can occur several years after treatment. Where little or no experience with managing this tumour exists is important to beware of its clinical behaviour and the treatment options hence this case report. We present unusual case of histologically confirmed malignant hemangiopericytoma of left thigh in 70-year female with metastasis to lungs with review of literature, to demonstrate the malignant character of this neoplasm and its variable prognosis.

Introduction

Hemangiopericytoma is rare tumour, first describe in 1942 by Stout and Murray in the lung. Comprising of capillary blood vessels and proliferating perivascular round cells, warranting hemangiopericytoma as a proper descriptive name [1].It has been defined as tumour composed of spindle to oval shaped undifferentiated cells, which proliferate around and are intimately associated with prominent, thin walled, often branching vessels. The tumour has been reported to occur in the lungs [1]. However it was not until1949, when Stout (15) reported 25 additional cases, that the tumour received widespread recognition. Extremities, pelvis [2], head and neck [3], back retro peritoneum and abdomen [4]. Primary central nerves system occurrence has also been reported [5]. Manifestation in bone is extremely rare but has been reported in afoot bone [6].

However, there are difficulties in the histological diagnosis of hemangiopericytoma because other soft tissue neoplasms may have areas of rish hemangiopericytoma -like vascularity [16,17]. It occurs commonly in fourth to fifth decade of life and show no sex predilection [18]. Surgical excision is the stander of care as HPC have a high recurrence rate as well as 20-30% of case may show malignant course [7]. Chemotherapy for unrespectable and disseminated HPC can lead to complete or partial remission of the disease in 50% of the cases[7]. There is no consensus on the type of chemotherapy treatment to employ, but by analogy with other forms of soft tissue sarcoma, doxorubicin +ifosfamide -based regimen is usually the first choice.

Enzinger and Smith [8] have suggested that malignancy can be predicted from a large tumour size, presence of necrosis, increased cellularity and over 4 mitoses per high power filed. However, clinical tumour behaviour and the occurrence of distance metastasis may be only way to conform the malignant nature of tumour. In the series of Enzinger and Smith, local recurrence precede metastasis in 11 of 17 patients who developed metastasis [19]. The lungs and bone are the most common sites for metastasis as occurred in this patient[19,20].

Case Report

A70 year old female patient was seen in April 2019 with 10 years painless left thigh swelling with slow growing, within month before medical seek the mass increase in size and limitation leg movement and complain pain but no pyrexia, there was no weight loss. Examination revealed healthy female patient with 10cm by 8 cm swelling on the posteromedial aspect of the left upper thigh(Figure1). It was firm, non-tender and non-pulsatile. Overlying skin was not worm and there was no significant inguinal lymphadenopathy. Past history, patient known case of diabetic mellitus (DM) since 25years under regular treatment metformin 500mg x bid.

Family history unremarkable. Radiological examination of affected leg by CT -scan 29/12/2018 show large mass from medical aspect of thigh that extend up word to suprapubic region causing lateral deviation of vagina. Metastatic work up done, no distance metastasis. 23/1/2019 True cut biopsy was don, histopathology shows malignant soft tissue sarcoma as cellular connective tissue neoplasm comprising proliferative spindle cells, immune stains done which show. CD 34 Strong positive and CD99, BCL2 and Vimentinpositive, EMA patchy focally positive while SMA and desmin are negative so immunohistochemistry in keeping with Malignant Hemangiopericytoma (Figure 2).

Refer for surgery but surgeon refuse, then chemotherapy start 27/1/2019 with ifosfamid 1500mg /m2 IVinf d1-4 + doxorubicin 20mg/m2 iv continues infusion on d1-3. Every 21days for three cycles than evaluation done by CT scan of abdomen + left thigh with oral and iv contrast in 17/4/2019 show large enhancing soft tissue mass lesion in the left upper thigh medially localized to the subcutaneous fat and pushing the perineum to right side, extending superiorly to the subcutaneous fat anterior to the symphysis without infiltrating of adjacent muscles, measuring about 8x10x11cm in TSxAPxCC dimeters.

Figure 1: Case of hemangiopericytoma.

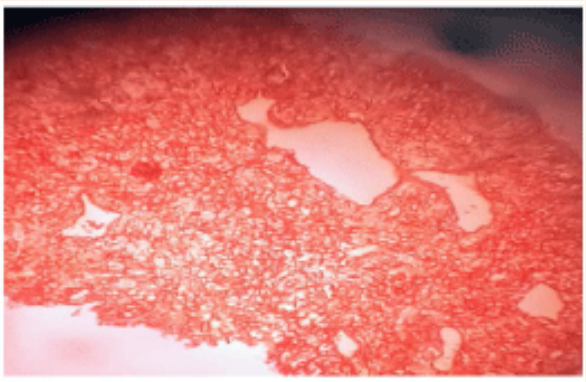

Figure 2: Cellular soft tissue tumor with staghorn blood vessels tumor cells are strong positive for CD34 in keeping with hemangiopericytoma .CD 99. TLE1, Pankeratin, S100 and bcl 2 all negative (not Shown).

Abdominopelvic CT-Scan, normal, Continue three cycles chemotherapy with same protocol,6cycle complete in 13/7/12019 evaluation done by CT-SCAN 29/7/2019 (multi-slice left femur CT scan with iv contrast show minimal progressive course of left upper thigh suspicious soft tissue sarcoma as regard size & extension (11x70x90mm) AP, TV &CC respectively, no evidence of nearby main vascular or intra-muscular or intra-osseous invasion, no calcification. patient developed DVT put on anticoagulation and supportive treatment.

In 11/112019, CT scan done abdomen-pelvic no significant change. CT scan Left thigh show left hyper-vascular anterior upper thigh highly suspicious soft tissue sarcoma (120x80x100mm). CT scan chest show right lung peripheral solitary nodule <5mm without calcification / metastasis, left lung pneumonia, multiple small mediastinal lymph nodes. 11/12/2019 start palliative chemotherapy with cyclophosphamide oral tab 100mg po daily for 21days every 28days first cycle, patient complete 3cycles with stable her condition till date of publish this case.

Discussion

The clinical behaviour of hemangiopericytoma predication is considerably difficult. This may be because it is heterogeneous entity with specific component variant that are yet to be. Identification of such variant will make management decision easier.Clinically, the hemangiopericytoma usually present as painless enlarging mass.in contrast its possible counterpart, the benign glomus tumour, of which the solitary group is the most common type, is usually located on an extremity and charlatanically present as a small painful mass(Table 1) lists helpful features in the differentiation of the benign solitary glomus tumour from the hemangiopericytomas.

Hemangiopericytoma is highly vascular tumour and occasionally may cause clinically significant arteriovenous shunting. With Doppler sonography, from the experience of others [21], it is likely that arteriovenous shunting would be shown in hyper vascularity neoplasm such as hemangiopericytoma.Wide surgical excision is the treatment of choice [9-11]. The unpredictable malignant behaviour of tumour, adjuvant therapy is desirable. Hemangiopericytoma is poorly radiosensitive but radiotherapy has been used with some success[9,12]. Primary and metastatic tumours are Chemosenstive[9]. Chemotherapy with methotaxate, actinomycin D, Cyclophosamide and vincristine in various combinations.

Table 1: Differential features of solitary Glomus VS. Hemangiopericytoma Tumor.

	Solitary Glomus	Hemangiopericytoma
Pain	Usually sever paroxysmal	Usually absent
size	Small (1-5mm)	Varies -usually large
Anatomical location	Skin, subcutaneous tissues hand, feet& often of nail beds (less common viscera)	All anatomical sites of body
Vascular Element	Arterial side vascular system	Capillary or venous side vascular system
Malignant potential	Absent	Approximately 50%
Light Microscopy	Polyhedral cells large round or ovoid nuclei & clear cytoplasm	Sheets of ovoid or spindle cell with indistinct cytoplasm &large various size nuclei
	Tumor cells separated by hyalinized eosinophilic stroma	Tumor cells around small vascular spaces
	No mitosis	Mitosis may be seen
Electron microscopy	Two cells: polyhedral smooth muscles cell & mast cell with granulas	Cells of various shapes and sizes with no basement membrane
	Large cytoplasm with number of filaments	These cells separated by basement membrane of endothelial cells
	Mitochondria are round or ovoid	Mitochondria rod-shaped or ovoid

Has also been used with success [9]. Combined radiotherapy and chemotherapy are to be recommended for metastasis, recurrence and inadequate excision. Patient should be followed up regularly because of local and distant tumour recurrence[13]. pulmonary metastasis can be fatal due to complication pneumothorax and hemothorax [14]. As with other soft tissue sarcomas, modern imaging techniques allow pre-operative assessment of the size and location of hemangiopericytoma, the degree of involvement of surrounding structures, and the vascularity of tumour[22]. Hemangiopericytoma should be consider in the differential diagnosis of hyper vascularity soft tissue mass arising in the retro peritoneum, pelvis or lower extremity. [22].

Conclusion

Hemangiopericytoma is slow growing but malignant tumour with capacity for local invasion and spread through the bloodstream. There is no single histologic criterion which will reliably differentiate the malignant from benign hemangiopericytoma. Thistumour like other soft tissue sarcomas, is difficult to control at their primary site by limited local excision. High recurrence rate result when simple emulation or excision is performed on the basis that the tumour appears to be encapsulated. Wide excision which may include amputation in selected cases is treatment of choice. Metastatic lesion, particularly in the lung, should be evaluated and resected whenever feasible. Radiation therapy may covert a non-respectable lesion into respectable one and provide significant palliation. The usefulness of irradiation therapy in management of this neoplasm is not well defined. Published reports of it use are few and this may be due to general consensus that most soft tissue sarcomas are radio resistant. Survival is based on a life time clinical course rather than five -year period because this tumour may grow slowly and recurrence may still develop after along interval.

References

1. Stout AP, Murray MR (1942) Hemangiopericytoma: Avascular tumour featuring Zimmermann's pericytes. AnnSurg 116:26-33.

2. Ahmad GF, Athavale R, Hamid BN. Davies HumphreysJ (2004) Pelivc malignant hemangiopericytoma mimicking an ovarian neoplasm; a case report. J Repred Med 49:404-407.

3. Pitluk HC, Conn Alhassan MB, Kyari Mohammed A (2003) Hemangiopericytoma of eyelid and orbit: report of 2 cases. Nigerian Journal of Surgical Research 5: 166-170.

4. J Jr (1979) Hemangiopericytoma. Literature review and clinical presentation. Am J Surg 137:413-416.

5. Spatola C, Privitera G (2004)Recurrent intracranial hemangiopericytoma with extracranial and unusual multiple metastases: case report and review of the literature. Tumori 90: 265-268.

6. Derouin MR, Key JJ, Caminear DS, Sella (2004) Hemangiopericytoma of the navicular: case report. J Foot Ankle Surg 43: 191-194.

7. T AKman, Alacacioglu, D Dolek, T Unek, D Gurel, et al. (2014)Malign recurrence of primary chest wall hemangiopericytoma in the lung after four years: a case report and review of literature. Case Rep Oncol Med 470268.

8. Enzinger FM, Smith BH (1976) Hemangiopericytoma: an analysis of 106 cases. Human Pathol7: 61-62.

9. Atkinson JB, Manhour GH. Isaac H, Ortega JA (1984)Hemangiopericytoma in infant and children. A report of six children. Am J Surg 148:372-374.

10. Grener RE, Moor GE, Pickern JW (1974)Hemangiopericytoma. Am Surg 179:128-132.

11. Perugia D, Basil A, Massoni C, Barletta V (1999)Hemangiopericytoma in the distal third of the arm. Int Orthop 23:184-186.

12. Friedman M, Egan JW (1960) Irradiation of hemangiopericytoma of Stout. Radiology 74: 721-729.

13. Yoshida J, Nakano T, Akao M, Tanimura A (1998) Pneumothorax due to hemangiopericytoma metastasis from thigh. JPn J Thorac Cardiovasc Surg 46:523-525.

14. Cheng YL, Yu CP, Hsu SH, Lee SC (2000) Hemangiopericytoma of the pleura causing massive hemothorax. J Formos Med Assoc 99: 428-430.

15. Lorigan J G, David C L, Evans H L, Wallace S (1989) The clinical and radiologic manifestation of hemangiopericytoma. Am J Roentgenol 153: 345-349.

16. Enterline H T (1981) Histopathology of sarcomas. Semin Oncol 8: 133-155.

17. Paullada J J, Lisci Gaemilla A, Gonzales Angulo, Jurado Mendoza J, Quijano Narezo M et al. (1968) Hemangiopericytoma associated with hypoglycemia. Metabolic and electron microscopic studies of case. Am J Med 44: 990-999.

18. Gitelis S Ryan W G, Rosenberg A G, Templeton AC (1986) Adult -onset hypophosphatasemia ostomalica secondary to neoplasm. J Bone Joint Surg Amer 68: 134-138.

19. Goldman S M, Davison AJ, Neal J (1988) Retroperitoneal and pelvic hemangiopericytoma; clinical radiologic and pathologic correlation. Radiology 168:13-17.

20. Feldman Seaman W B (1964) Primary thoracic hemangiopericytoma. Radiology 182:998-1008.

21. Taylor K J, Ramos I, Carter D, Morse SS (1988) Snower D, Fortune Correlation of Doppler US tumor signals with neovascular morphologic features. Radiology 166: 57-62.

22. Alpern M B, Thorsen M K, Kellman G M, Pojunas K, Lawson T L (1986) CT Appearance of Hemangiopericytoma. J Comput Assit Tomogr 10: 264-267.