METASTATIC HAEMANGIOPERICYTOMA: A RARE VARIETY OF SOFT TISSUE SARCOMA PRIMARILY INVOLVING THE LEFT GLUTEAL MUSCLES

Muhammad Younus Awan, Irfan Shukr*

01 Mountain Medical Battalion, *Combined Military Hospital Rawalpindi

INTRODUCTION

Haemangiopericytomas have been reported in almost all anatomic sites, but the tumours reported in the extremities or in the intramuscular locations with atypical histological features and malignant behaviour, are rare[1]. Tumours involving the extremities, pelvis, head and neck, back, retroperitoneum and abdomen have been reported previously[2,3,4]. Haemangiopericytoma are a variety of soft tissue sarcomas which are vascular in origin and consist of pericytes[5]. Malignancy in haemangiopericytoma is predicted from large tumour size, presence of necrosis, increased cellularity and over 4 mitosis per high power fields suggested by Enzinger and Smith[6]. However, tumour behaviour and the occurrence of distant metastasis may clinically be the only way to confirm the malignant nature of the tumour. Metastasis or local recurrence may take several years to develop[7].

CASE REPORT

A 77 year old male presented to surgical department, Combined Military Hospital Rawalpindi with a huge swelling over the left gluteal region for the past 30 years. Initially it was a small swelling about the size of a table tennis ball which the patient noticed after blunt trauma to the left gluteal region. It was not associated with pain, fever, night sweats and did not hamper the daily activities or movement of the patient. About two years ago, it rapidly increased in size and became painful. The pain was dull, continuous, non- radiating and aggravated by pressure over the swelling. Patient did not have history of any other swelling in the body. He also gave a history of 20 kg weight loss during 2 years and of chewing tobacco for the last 40 years.

On examination, there was a non tender, firm, hemispherical swelling measuring 30 cms in diameter over the left gluteal region. Its margins were ill defined, overlying skin was mobile with dilated superficial veins and temperature was slightly raised. It was fixed to underlying muscle, non- compressible, non-pulsatile, non- fluctuant, non- reducible and did not transilluminate. Distal neurovascular status was intact and there was no inguinal lymhadenopathy.

A provisional diagnosis of soft tissue sarcoma was made and the patient was admitted for further workup. Chest X Ray (PA view) showed multiple canon ball opacities in both lung fields (Fig 1). CT scan of the chest, abdomen and pelvis was done to see the relationship of the tumour with bone and to look for metastases. The tumor had no anatomical connection with the underlying pelvic bones. Nodules representing metastases were seen in the lungs (Fig 2). An incision biopsy was done and histopathological examination was carried out at Armed Forces Institute of Pathology, Rawalpindi. Microscopically, the tumour was composed of plump to spindle shaped cells arranged around blood vessels. Some areas had myxoid background. Tumour was strongly positive for CD 34. Cellularity and areas of necrosis were strongly in favour of malignant haemangiopericytoma.

Since the tumor had already metastasized to the lungs, surgery was not an appropriate treatment option. We referred the patient to the oncology team for opinion regarding chemo/ radiotherapy. Unfortunately, the patient was lost to follow up.

DISCUSSION

Malignant hemangiopericytoma arising from the pericytes are a rare soft tissue tumors of perivascular origin[1]. Literature shows that these tumours are more common in the lower

Correspondence: Maj Muhammad Younus Awan, Surgical Specialist, 01 Mountain Medical Battalion.
Email: younus01@hotmail.com
Received: 28 Feb 2011; Accepted: 02 Oct 2012

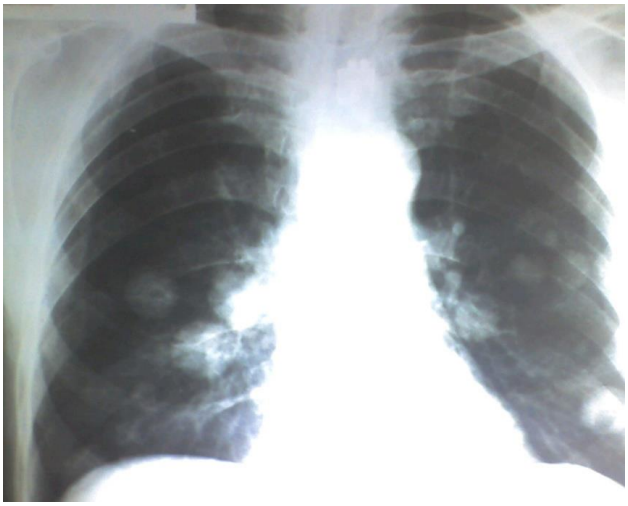

Fig. 1: Chest X ray showing multiple cannon ball opacities.

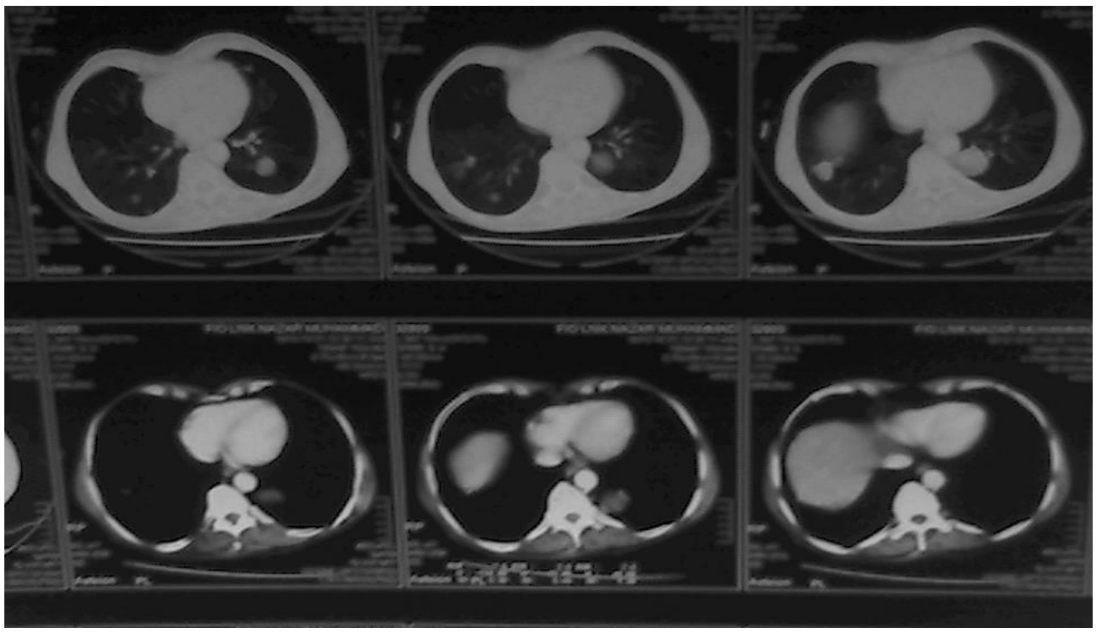

Fig. 2: CT scan showing nodules in the lungs.

extremities, followed by retroperitoneum and other sites[8]. Our patient had metastatic haemangiopericytoma involving the left gluteal region, which is in keeping with this observation. Clinically, they are slow growing tumours. Though the presentation is similar to soft tissue sarcomas, as in the case described, histopathological findings are charactesistic and clinch the diagnosis. On a microscopic level, they consist of tightly packed cells around thin-walled, endothelium-lined vascular channels ranging from capillary-sized vessels to large gaping sinusoidal spaces. Haemangiopericytomas are highly malignant tumors and generally metastasize to lungs, bones, liver, and lymph nodes, with rates ranging from 11.7% to 56.5%.[8] Pulmonary metastases are usually considered fatal due to the pneumothorax and haemothorax complicating them[9,10].

Surgery is the main treatment modality for soft tissue sarcomas but combined radi-chemotherapy is recommended for metastatic disease as well as recurrent and initially inadequately excised tumours[7]. Chemotherapy and radiotherapy are often required for the treatment of malignant hemangiopericytomas[11]. Both the primary tumour and metastasis are chemosensitive; methotrexate, actinomycin D,

cyclophosphamide and vincristine have been used with some success in various combinations[12]. Sensitivity to radiotherapy remains poor, though this treatment modality is also used in some cases[13].

CONCLUSION

In patients presenting with painless slow growing lumps over the extremities, haemangiopericytoma should be considered as a possible etiology. Every effort should be made to confirm the diagnosis timely since surgery can be curative in early disease.

REFERENCES

1. Zanelli M, Andreini L, Galanti G, Cortecchia S, Nannini R, Bondi A. Hemangiopericytoma-solitary fibrous tumour of soft tissue: description of a case showing atypical histological features. Pathologica 2010; 102(1):30-2.
2. Ahmad GF, Athavale R, Hamid BN, Davies-Humphreys J. Pelvic malignant hemangiopericytoma mimicking an ovarian neoplasm; a case report. J Reprod Med 2004; 49:404–7.
3. Alhassan MB, Kyari F, Mohammed A. Haemangiopericytoma of the eyelid and orbit: report of 2 cases. Nigerian Journal of Surgical Research 2003;5:166–170.
4. Pitluk HC, Conn J. Jr Hemangiopericytoma. Literature review and clinical presentations. Am J Surg1979;137:413–6.
5. Hansen T, Gaumann A, Ghalibafian M, Hoferlin A, Heintz A, Kirkpatrick CJ. Haemangiopericytoma of the thyroid gland in combination with Hashimoto's disease. Virchows Arch 2004; 445 (3): 315-9.
6. Enzinger FM, Smith BH. Haemangiopericytoma: an analysis of 106 cases. Human Pathol 1976;7:61–2.
7. Thanni LO. Extremity haemangiopericytoma, a case report from Nigeria.Afr Health Sci 2005; 5 (3): 261-4.
8. Sherwani RK, Rahman K, Akhtar K, Siddiqui FA, Varshney BK, Sherwani M. Malignant hemangiopericytoma in the gluteal region: A rare entity. Indian J PatholMicrobiol 2008; 51: 39-41.
9. Yoshida J, Nakano T, Akao M, Tanimura A. Pneumothorax due to hemangiopericytoma metastasis from the thigh. Jpn J Thorac Cardiovasc Surg 1998; 46: 523–5.
10. Cheng YL, Yu CP, Hsu SH, Lee SC. Hemangiopericytoma of the pleura causing massive hemothorax. J Formos Med Assoc 2000; 99: 428–30.
11. Dube VE, Parlson JF. Metastatic hemangiopericytoma cured by radiotherapy. J Bone Joint Surg 1974; 56A: 833.
12. Atkinson JB, Manhour GH, Isaacs H, Ortega JA. Haemangiopericytoma in infants and children. A report of six children. Am J Surg 1984;148:372-4.
13. Friedman M, Egan JW. Irradiation of haemangiopericytoma of Stout. Radiology 1960; 74: 721–9.