

Recurrent Angiosarcoma on the Back in a Young Male: A Case Report

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Abstract

Angiosarcoma is a malignant disease of inner lining of the blood vessel which can occur at any age and anywhere in the body like skin, liver, breast, bone, soft tissues cardiac tissues and after radiotherapy or from chronic lymphatic obstruction. It usually occurs between 60-70 years of age¹. In this case, it occurred in a young male, which was previously excised but recurrence occurred and presented as a lump of 20×15 cm, which was non tender, gradually increasing in size, firm in consistency however other systemic findings were unremarkable. Haematological work up was done which was within normal limits. Ultrasound Doppler of swelling showed multiple vascular channels with fat and soft tissue suggestive of a haemangioma. Excision and biopsy were done under general anaesthesia and histopathology report showed fibro-fatty tissue with vascular malformation. In view of recurrence it suggested angiosarcoma, which is treated by surgery followed by chemotherapy and its definitive diagnosis can only be made by biopsy.

Keywords: Hemangiosarcoma, recurrence, biopsy.

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Introduction

Angiosarcoma is the tumour of tunica intima of the blood vessels. It can occur anywhere in the body like skin, breast, liver, spleen, and deep tissues. Prognosis of angiosarcoma depends on many risk factors including: age, gender, site, size, surgery implicated¹ tumour size less than 5cm carries a good prognosis. Angiosarcoma occurring in the heart have poor prognosis and high chance of recurrence, increasing chances of destruction if diagnosed late. As it rarely occurs in the heart, commonly occurring in the right atrium, and have a poor response to radio-chemotherapy; surgical treatment can also be fatal^{2,4,5}. Angiosarcomas occurring in the skin are metastatic lesions, which re-

spond less to therapy and carry a fatal diagnosis. Angiosarcomas in face and scalp carry poor prognosis because when they are diagnosed, they are of larger size tumour that are high grade and usually metastasize before the diagnosis³. Most of the time the clinical presentation is different, thus early diagnosis is the only possible way for good prognosis because it measures early³. It presents with symptoms when the disease is at its advance stage or according to clinical area from where it is arising, either mass, bleeding, fracture or terminal stage disease features. Breast angiosarcomas are rare and occur in young women who have previously been treated (chemo radiations) for breast carcinoma and are also difficult to treat⁶. Angiosarcoma can present as a soft lump, infection or a bruise. Various investigations are available but a definitive diagnosis is made after biopsy. Surgery is the main stay of treatment and then chemotherapeutic regimen can be given, which includes mesna, doxorubicin and ifosfamide, and paclitaxel and docetaxel are effective in angiosarcoma of head, neck and scalp.

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Case Report

A 20-year old Asian male came to the hospital with the complain of recurrent benign swelling at the back for 4 years. According to the patient, he was all right in July 2014, when he noticed a swelling in his back on the right side of midline, which was gradually increasing in size and shape. Swelling was not associated with pain, fever and nausea or vomiting. The swelling was operated on 23rd July 2015 and was about 20×15 cm in size. Histopathology report revealed that it was a haemangioliopoma. On 12th February 2019 the patient was readmitted through the OPD with complains of swelling on the right side of midline, which was gradually increasing in size and not associated with pain.

He had no past medical history but was hospitalized for 5 days and operated for the same complain in 2015. His family history was not significant for any tumour, hepatitis B, hepatitis C. There was no history of tuberculosis contact. No history of previous or active addiction nor any history of blood transfusion was present. He did not receive any systemic treatment and had no family history of tuberous sclerosis complex or any other inherited conditions.

Upon general examination, he was a young-looking male having thin and lean built lying comfortably on bed with blood pressure measuring 120/70 mm Hg, pulse 100 bpm, respiratory rate 20 bpm and temperature 98-degree Fahrenheit. No signs of oedema, jaundice, cyanosis or clubbing were present. Abdomen was soft, non-tender and chest was clear. On cardiovascular examination, both S1+S2 were audible with no added sounds and thyroid examination was normal. Other systemic examination was unremarkable.

On local examination scar mark was present in the midline of the posterior side of chest, 2 swellings present over the medial border of scapula extending up to 6th-7th rib on the back. First swelling was about 8×7 cm in size, firm in consistency, immobile in nature, irreducible, slip sign negative

and on auscultation, there was not any bruit appreciated. Second swelling was about 6×5 cm in size, firm in consistency, immobile in nature, irreducible, adherent to the underlying structures, slip sign negative and on auscultation there was not any bruit appreciated.

At presentation his Hb was 13.6 mg/dl, TLC 6.3 10³ u/l, platelets 184 10³ u/l, urea 22 mg/dl, and creatinine 0.9 mg/dl, Hbs Ag and Anti Hcv were non-reactive. Urine d/r showed 0-1 pus cells.

Ultrasound Doppler of swelling showed multiple lobulated lesions at the right side of the chest showing significant vascular channels along with fat and soft tissue suggestive of haemangioma.

Patient and family members were counselled for surgery and related risks on 14th February 2019. Written and informed consent was taken. One pint of whole blood was arranged. Patient was prepared for surgery, he was kept nil per oral, tablet laxtonil 3 mg was given 12 hours before surgery. Pre-operative blood pressure was recorded as 140/100 mmHg, pulse 70 bpm, respiratory rate 20 bpm and fasting blood sugar 91 mg/dl. Prophylactic injection Augmentin 1.2 gm was given at the operating table. Injection ringer lactate 1000cc I/V stat was given to preload the patient.

Under General anaesthesia, After all aseptic measured an elliptical skin incision given on previous scar. Fat was separated and then the mass separated from surrounding structures. During separation multiple bleeders were present and due to excessive bleeding whole blood was attached during surgery. Haemostasis was secured with chromic 0. Rediwig drain was placed and fat was closed with vicryl 4.0. The skin was closed with prolene 2.0. Aseptic dressing was applied. No post-operative complications were noted.

The mass was of 7×7 cm, unencapsulated, adherent to underlying structures and another 5×5 cm unencapsulated mass was adherent to the underlying structures with multiple bleeders.

Histopathology report showed fibro-fatty tissue of varying size and shape consistent with vascular malformations. However, multiple foci of new blood vessels/capillaries formation were noted. These were crowded with some pleomorphism. The nuclei were prominent and few mitosis noted (upto 3/10 HPF). In view of recurrence, a possibility of angiosarcoma could not be excluded.

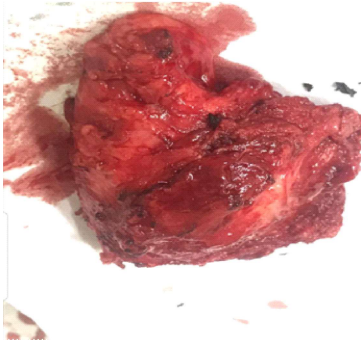


Fig 1. Gross: Angiosarcoma on back

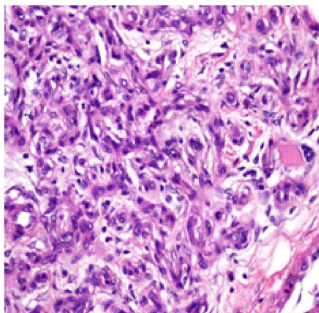


Fig 1.1. Histopathology slide of angiosarcoma on back

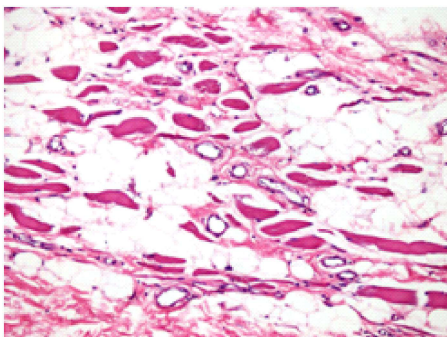


Fig 1.2. Histopathology slide of angiosarcoma on back

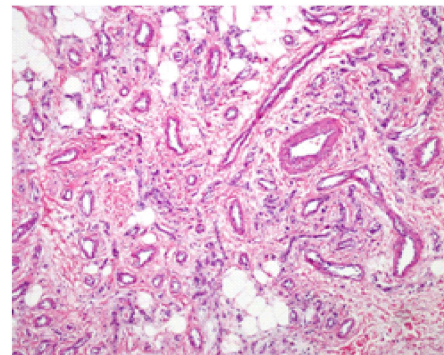


Fig 1.3. Histopathology slide of angiosarcoma on back

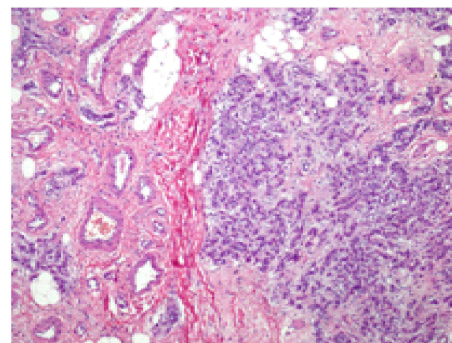


Fig 1.4. Histopathology slide of angiosarcoma on back

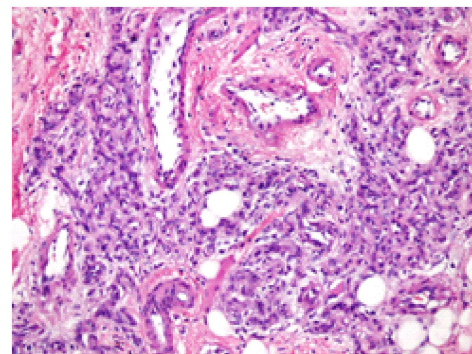


Fig 1.5. Histopathology slide of angiosarcoma on back

Discussion

Angiosarcoma is a rare, metastatic disease that may be idiopathic or due to chronically obstructing lymphatics but in this case, it was fat cells along with blood vessels. Angiosarcoma occurs at the same rate in both men and women but

at advance stage. Clinically it presents as a soft gradually enlarging mass which is most commonly tender with symptoms of anaemia in patients but sometimes presents as a bruise, oedema or non-healing ulcer. Prognosis of angiosarcoma depends on the age of the person, gender (like in females there are more chances), on which site it is arising, size of the tumour, which surgery is going to be implicated, chances of recurrence after surgery and response of adjuvant therapy on the tumour³. In this case, it was non-tender, covered by skin with no signs of inflammation except the gradually enlarging mass. There are two types of vascular sarcoma, hemangiosarcoma (arising from the cells that make the wall of blood vessels) and lymphangiosarcoma (arising from the cells that make up the wall of lymphatic vessel), and alveolar soft part sarcoma (arising from the cells that make up the connective tissues).

Clinically cutaneous angiosarcoma is most common and most commonly occurs on the face and scalp, where it is highly spreadable locally and has a poor prognosis^{1,7} but in this case it was present on the back of chest where it occurred just beneath the skin hence called subcutaneous angiosarcoma. The angiosarcoma, which occurs in deep tissues like the liver, is called hemangioendothelioma. Due to soft tissue involvement MRI is more reliable than CT scan and we can assess and differentiate the preoperative changes regarding the size of the tumour and its expandability. Biopsy is the diagnostic test, which shows atypical cells with large vascular spaces and mitosis. Radiation followed by resection of tumour is the mainstay of treatment for localized disease. However, surgical resection is contraindicated in metastatic disease, but in this case both times surgical resection was done followed by chemotherapy.

Follow-up after treatment should be done every 3 months in first 2 years and then every 6 months for the next 3 years and then after 5 years, follow-up should be done annually.

Conflict of Interests

Authors have no conflict of interests and received no grant/funding from any organization.

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