

Giant Haemangiopericytoma of the Uterus

Dam Purvita · Saha Sudip Kumar ·
Banerjee Sharmistha · Ganguly Sourav

Received: 7 April 2008 / Accepted: 10 June 2010 / Published online: 7 March 2012
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Introduction

Hemangiopericytoma is an uncommon mesenchymal neoplasm which may originate anywhere in the body. It is thought to arise from the pericytes, contractile spindle cells that surround the capillaries and postcapillary venules [1]. Tumors usually have uniform elongated cells surrounded by branching network of thin walled vessels of various sizes and shapes. Good survival is common in hemangiopericytoma patients treated with curative intent. However, local and distant recurrences may occur after a prolonged disease-free interval, emphasizing the need for long-term follow-up. Prognosis of hemangiopericytoma of uterus is better [2]. Primary treatment involves radical hysterectomy with bilateral salpingo-oophorectomy. Till date the biggest reported uterine tumor was by Trace Cornforth in 2003 in ? about.com women's health was at around 140 pounds [3].

Case Report

AB, a 46 years old nullipara, widow attended the Gynae OPD of Medical College, Kolkata with history of generalized swelling of abdomen for 20 years which increased massively in the last 5 years along with swelling of lower limbs on 4 October 2007. She also had respiratory distress for last 2 months. Menstrual history was regular. On examination G.C. was fair, abdomen was hugely distended with a mass extending from xiphisternum to symphysis pubis and flanks full. Abdominal veins were engorged. The mass was solid in nature. Her weight was 80 kg. A per vaginal examination only revealed flushed cervix and uterus or adnexae could not be made out separately. A CT scan on 5 October 2007 revealed huge ovarian cyst with calcification and minimal ascites. There were areas of low attenuation too. CA-125 was 48. Other investigations were within normal limits. Long history and good general condition suggesting a benign mass, decision for laparotomy was taken. On 11 November 2007, her abdomen was opened by midline incision extending from umbilicus to suprapubic region. A huge solid to cystic mass of 20 × 25-cm with engorged veins was noted. On gradually lysing filmy peritoneal and omental adhesions, the mass was well visualised arising from the fundus of uterus. The uterus was approximately 8 weeks in size. The entire mass with uterus was delivered out and bilateral apparently healthy tubes and ovaries were observed. Provisional diagnosis on laparotomy was huge fibroid uterus. Ureters were identified and total abdominal hysterectomy and

Dam P., RMO cum clinical Tutor · Saha
S. K., Associate Professor · Banerjee S., MD PGT ·
Ganguly S., Rotating Internee
Department of OBGYN, Medical College and Hospital,
Kolkata, India

Dam P. (✉), RMO cum clinical Tutor
26 Sarat Chatterjee Road, Kolkata 700089, India
e-mail: purvita_mdgo@yahoo.com

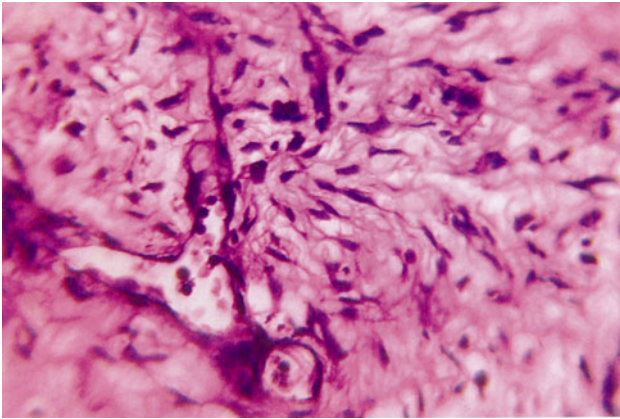


Fig. 1 Microphotograph showing focal edematous area surrounding network of thin walled vessels (H&F, ×400)

bilateral salpingo-oophorectomy was done. Surprisingly the mass weighed 20 kg. Postoperative recovery was uneventful. Before discharge her weight was 58 kg. Histopathology was even more surprising. There were focal oedematous areas with elongated cells surrounding branching network of thin walled vessels, suggestive of haemangiopericytoma (Fig. 1).

Discussion

Haemangiopericytoma account for less than 2% of soft tissue sarcomas [4]. In a series of 106 patients reported by Enzinger and Smith, 25% were located in the pelvis [5]. It may occur at any age, but is most common in the forties, fifties and sixties [6]. Our patient was in her fourth decade. Haemangiopericytoma usually presents as a painless mass,

but symptoms following pressure to adjacent viscera may occur as seen in our patient. Various paraneoplastic reactions may be anticipated with haemangiopericytoma, but were not present in our patient. Areas of low attenuation in CT possibly signify zones of necrosis. It is a highly vascular tumor at times with significant arteriovenous shunting; pelvic tumors in particular may be very large when first seen leading to excessive hemorrhage at surgery which was fortunately absent in our case [7]. What makes our case unique is the weight of the tumor, 20 kg, as after a lot of net browsing we have not been able to locate a bigger or heavier haemangiopericytoma. Whenever there is a hypervascular soft tissue mass arising from retroperitoneum or pelvis, one must consider hemangiopericytoma as a differential diagnosis.

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