Giant retroperitoneal desmoid tumour

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Abstract

A giant retroperitoneal desmoid (30 × 15 cm), in a 16-year old girl arising from psoas fascia is reported. Despite debulking surgery, adjuvant radiotherapy, anti-oestrogen agents and non-steroidal anti-inflammatory agents, 3 years later she died from tumour invasion of major blood vessels and bowel, leading to massive gastrointestinal bleeding.

Case report

A 16-year old girl presented with symptoms suggestive of left pyelonephritis which turned out to be a left-sided large non-tender abdominal mass obstructing the left kidney. A CT scan showed a 30 cm × 15 cm retroperitoneal soft tissue mass with necrotic areas, extending from the left kidney up to the level of the urinary bladder. After a preliminary core needle biopsy debulking excision of the mass was done with a left nephrectomy. Histology confirmed aggressive fibromatosis (Figure 1).

Figure 1. Desmoid tumour composed of elongated, slender spindle shaped cells arranged as sweeping fascicles (× 10, H & E).

Post-operatively she was treated with tamoxifen and sulindac for the residual tumour.

With evidence of residual mass a course of radiotherapy was added. She was lost for follow up for 3 years, and presented with an abdominal mass, bilateral ankle oedema and flexion deformity of the hip, due to contraction of the ipsilateral ileo-psoas muscle. Repeat CT scan revealed residual tumour growth extending to the stomach and engulfing midline structures such as aorta, mesenteric arteries and inferior vena cava, making it inoperable (Figure 2). Her general health deteriorated rapidly and she died following a sudden episode of haematemesis and melaena.

Desmoid tumours (aggressive or musculo-aponeurotic fibromatosis) first described in 1832, are rare slow growing, histologically benign tumours with no metastatic potential. Out of three clinicopathological variants based on location, abdominal desmoids account for half the cases [1]. Majority arise in the musculo-aponeurotic structures of the anterior abdominal wall, in women during or following pregnancy. Intra-abdominal desmoids tend to occur in the mesentery or pelvic wall, often in patients having Gardner syndrome [2]. Giant retroperitoneal desmoids are rare and there are no documented cases of giant retroperitoneal desmoids arising from psoas fascia, in the contemporary English literature. Despite their benign appearance they are locally aggressive leading to involvement of surrounding structures and recurrence even after wide local excision. Desmoids may occur at any age but are most frequent in the second to fourth decades. Molecular studies have demonstrated that the desmoids resemble neoplasms and form as a result of a clonal process [3]. The association with Gardner syndrome suggests genetic influences [2].

Wide local excision with clear margins in all directions is the therapy of choice, though it may not prevent local recurrence [1,4]. Although its efficacy remains controversial radiotherapy is recommended when negative resection margins cannot be obtained [5]. There are reports of tumour regression with the use of anti oestrogen therapy and non steroidal anti inflammatory drugs such as sulindac or indomethacin [6]. The use of combination chemotherapy regimens is under investigation [7].

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Figure 2. Contrast CT scan demonstrating the residual tumour as a large, ill defined, mixed density mass engulfing midline structures (A), extending paravertebally in to bony pelvis (B).

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Condoms and the Vatican

Pope Benedict XVI certainly needs to receive some good medical advice. Last June, in a speech to the Bishops of Botswana, Lesotho, Namibia, South Africa, and Swaziland, he said that "the traditional teaching of the Church has proven to be the only failsafe way to prevent the spread of HIV/AIDS". This is palpably untrue. The Catholic Church also needs to rein in officials, like Cardinal Alfonso Lopez Trujillo, who argue that HIV is small enough to pass through a condom. Such scientific untruths give the Catholic Church a bad name.

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