Giant desmoid tumor in the posterior abdominal wall/retroperitoneum

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ABSTRACT

Desmoid tumor is a rare soft tissue neoplasm arising from musculo-aponeurotic tissues constituting 0.03% of all neoplasms with a high rate of local recurrence after surgery. Radiotherapy may be used to prevent local recurrence. We report a giant desmoid tumour arising from the posterior abdominal wall / retroperitoneum.

Key words: Aggressive fibromatoses, desmoid tumor, radiotherapy, surgery

INTRODUCTION

The term “desmoid” was coined by Muller\(^1\) in 1838 and is derived from the Greek word “desmos”, which means “tendon-like”. Desmoid tumors also known as aggressive fibromatoses, are soft tissue neoplasms arising from musculo-aponeurotic tissues. These tumors are rare,\(^1,2\) constituting 0.03% of all neoplasms.\(^3\) We report a giant desmoid tumor arising from the posterior abdominal wall / retroperitoneum.

CASE REPORT

A 17-year-old female patient presented with a swelling over the right side of her back of four years’ duration. The swelling was insidious in onset and progressed slowly, but there was a sudden increase in size of the swelling over the last six months causing disability and disfigurement [Figure 1]. Local examination revealed a fixed, nontender, bosselated 36 × 29 cm soft tissue mass with varying consistency, longitudinally extending from the inferior border of the right scapula to the iliac crest and transversely extending from the mid-axillary line to the posterior mid-line. Dilated veins were visible over the mass. The mass was fixed to the chest wall and posterior abdominal wall but not fixed to the skin. Abdominal examination revealed the same mass extending to the retroperitoneum in the right hypochondrium and lumbar region. The right kidney was palpable and displaced anteromedially. The liver was not enlarged.

Abdominal ultrasound showed a large solid mass in the right posterior lumbar region with retroperitoneal extension causing displacement of the right kidney anteriorly. Duplex scan of the tumor did not detect any significant vascularity. Chest X-ray was normal. Fine needle aspiration cytology (FNAC) of the mass suggested features of peripheral nerve sheath tumour (Neurofibroma / Schwannoma). Computed tomography (CT) scan showed a large well-defined, heterogenous, enhancing soft tissue attenuation mass lesion in the retroperitoneum with extension to the back. The right kidney was displaced anteromedially. Inferior vena cava (IVC) was compressed. These features were consistent with a neurogenic tumour [Figure 2]. Magnetic resonance imaging (MRI) of the mass revealed a large heterogenous soft tissue lesion in the right retroperitoneal region extending from T\(_{10}\) to L\(_{1}\) level (20 × 14 cm). Superiorly, it was extending up to the level of the diaphragm displacing the right lobe of the liver anteriorly. The kidney was displaced anteriorly almost towards the mid-line. Ascending colon was shifted medially. IVC was compressed and displaced towards the mid-line. Medially, the tumor extended up to the right paravertebral region. Right psoas was normal.
Vertebral column, neural foramina and spinal canal were normal. These features suggested the possibility of a neurogenic tumor or rhabdomyoma/sarcoma.

The tumor was approached through right subcostal incision extending posteriorly and superiorly over the tumor. Anteriorly, the tumor was approached through a transperitoneal route. It was adherent to the right crus of the diaphragm and infiltrated into the muscles of the back and paravertebral region. The tumor was excised along with the right 12th rib and part of the muscles infiltrated by the tumor [Figure 3]. The diaphragmatic defect was repaired with polypropylene 1'0' suture in two layers after placing an intercostal drain. Posterior abdominal wall muscles were sutured and the defect was reinforced by a polypropylene mesh. The wound was closed in layers after placing a drain in the retroperitoneum. Postoperative period was uneventful.

The morphological appearance showed trabeculated yellowish to grayish glistening white areas and areas of hemorrhage. Histopathological examination showed benign tumor cells arranged in interlacing fascicles and bundles arranged in storiform pattern. Individual tumor cells had round, oval and a few wavy nuclei with indistinct eosinophilic cytoplasm. The stroma showed abundant myxoid area with cystic area. Features were suggestive of intraabdominal desmoid. The patient was referred for radiotherapy to prevent further recurrence since the tumor had infiltrated the muscles. The patient is alive with no evidence of disease at 18 months follow-up.

**DISCUSSION**

Desmoid tumors belong to a family of fibroblastic proliferations that include a variety of fibromatoses. More than 75% of these tumours arise in extraabdominal sites—primarily the shoulder girdle, inguinal area and lower extremities.[2] The cause of desmoid tumors is uncertain and may be related to trauma or hormonal factors. They may also have genetic association.[1,4] Abdominal wall desmoids occur most commonly in women of child-bearing age during or shortly following pregnancy—they may be associated with scars from prior operation or trauma. Intraabdominal sites account for less than 10% of all spontaneous desmoid tumors.[4] The tumors occur in 10-15% of patients with familial polyposis coli (Gardner’s syndrome). Desmoid tumors present often as a painless mass located deep in the abdominal wall arising from the rectus abdominis muscles or linea alba, distinguishable in most situations from hernias or intraabdominal masses by physical examination.

On microscopic examination, the tumors are composed of uniform, normal-appearing fibroblasts distributed through dense collagenous stroma. Nuclei are thin and elongated; mitotic figures are seen typically in less
than one per 50 high-power microscopic fields. The fibroblastic component has neither the cellularity nor pleomorphism required for classification of low-grade fibrosarcoma. The lesions are never encapsulated. They insinuate themselves along fascial planes and muscle bundles in a fashion identical to that of sarcomas. The natural history of abdominal wall desmoid tumors is one of local progression with potential invasion of intraabdominal structures, extremely rare pulmonary metastasis and multiple local recurrences after excision; 70-80% of recurrences develop within two years of excision.\[1,2\] The diagnosis of a desmoid tumor is confirmed by examination of a tru-cut core biopsy. If the lesion is less than 4 cm in diameter, excision biopsy is appropriate.

Wide local excision of the tumor involving excision of the full thickness of the abdominal wall is the operation of choice. Local recurrence rates are lower than for extraabdominal tumors, ranging from 10-40%.\[2\] Although the presence of a pseudo capsule may give the surgeon a false sense of security when excising a presumably benign mass, tumor cells invariably extend beyond the grossly apparent tumor, making marginal excision a seemingly inadequate operation. Recurrent tumors should be re-excised when the lesion can be resected grossly. Merchant et al.\[5\] concluded that attempts to achieve negative resection margins may result in unnecessary morbidity and may not prevent local recurrence. Operations that preserve function and structure should be the primary goal because the presence of residual disease has not been clearly shown to impact adversely on 5-year disease-free or overall survival.

Radiation therapy with doses of 5000-6500 centi-Gray (cGy) units is useful in controlling many unresectable tumors, producing complete responses. Response to radiation is often slow and desmoid tumors may require two years to shrink. Postoperative radiation should be considered only when there is grossly evident residual tumor following an attempt at resection. For marginal excision, careful follow-up and prompt excision of the recurrent tumor or radiation therapy is an effective alternative. A variety of hormonal, prostaglandin inhibitors and chemotherapeutic agents have been evaluated for the treatment of desmoid tumors when surgery and radiotherapy are no longer treatment options. Chemotherapy drugs used are vinblastine and methotrexate, doxorubicin, dacarbazine and carboplatin. Tamoxifen alone or in combination with indomethacin has shown a good response.\[1,2\] Nuyttens et al.\[6\] had conducted a Medline search using the keywords “desmoid”, “surgery” and “radiotherapy” and concluded that radiotherapy or surgery with radiotherapy (RT) results in significantly better local control than surgery (S) alone. Even after dividing the groups into cases with free and positive margins and cases with primary and recurrent tumors, the best local control is achieved with RT or S + RT. After surgery, MRI may be useful for monitoring recurrence.

This case is published because of its uncommon site and giant size. In young patients, whatever may be the tumor size, it is advisable to attempt excision of the tumor whenever possible hoping for better outcome as in our case which turned out to be a benign tumor. In our case due to extensive fibromatoses, excision was incomplete. The patient was treated with radiotherapy to prevent recurrence. However, long-term follow-up is required.

**REFERENCES**