CASE REPORT

Radical resection and improvised surgical reconstruction for a rare malignant triton tumour of intercostal nerve in a patient with neurofibromatosis type 1

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SUMMARY

A man aged 28 years, with neurofibromatosis type 1, presented with abdominal pain and visible right lower chest swelling. He had cutaneous neurofibromas and several café-au-lait spots. CT scan of the chest and abdomen revealed a giant mass most likely originating from the right 7th intercostal nerve, extending downwards into the abdomen causing massive hepatic compression. A Trucut biopsy showed malignant peripheral nerve sheath tumour. Radical surgical resection with wide margins was performed. The chest wall and diaphragm were reconstructed with improvised surgical technique with excellent results. Final histopathology report revealed a triton tumour of the intercostal nerve.

showed normal gall bladder and a large mass compressing the liver. CT scan of the chest revealed a well-defined complex right lower chest wall mass involving the diaphragm and extending downwards into the right upper abdomen causing massive liver compression ([figure 1A](#)). Haematological examination revealed that haemoglobin, white cell count and renal panel were normal.

Liver enzymes were markedly raised.

INVESTIGATIONS

- ▶ Chest X-ray
- ▶ CT scan chest
- ▶ Ultrasound scan abdomen
- ▶ Liver function tests
- ▶ CT-guided fine needle aspiration
- ▶ Pulmonary function tests

DIFFERENTIAL DIAGNOSIS

- ▶ Chest wall soft tissue sarcoma
- ▶ Neurofibroma
- ▶ Chondrosarcoma
- ▶ Malignant nerve sheath tumour

TREATMENT

Multidisciplinary tumour board's decision was to proceed for surgical resection.

Surgical approach was a right thoracoabdominal incision starting from the seventh intercostal space extending into the abdomen's right subcostal area. A large tumour arising from the seventh intercostal nerve was seen, involving a part of the diaphragm mainly extending into the abdomen and causing massive liver compression without any direct invasion of the hepatic parenchyma. Tumour was excised en bloc with sixth, seventh, eighth and ninth ribs, along with part of the diaphragm and abdominal wall muscles. The abdominal wall defect was repaired with a biological mesh. The chest wall was reconstructed with a methyl methacrylate Marlex mesh sandwich (MMS) plate. MMS was made and moulded according to the defect and contour of the chest wall. Holes were drilled in the cement plate corners and the surrounding bones. Additional six holes were made in the plate each 1 cm apart and #2 silk stitches were passed through the diaphragm and then through the holes in MMS and tied the latter on the outside as shown in [figure 2](#). The MMS plate was anchored by interrupted 2/0 prolene (Ethicon, Somerville,

BACKGROUND

Neurofibromatosis type 1 (NF1) also known as von Recklinghausen disease was first described by German pathologist Friedrich Daniel von Recklinghausen in 1882. This is a rare genetic disorder which equally affects both genders, all the races and ethnic groups. The incidence is ~1 in 4000 births.¹ Malignant triton tumour (MTT) is a rare subset of malignant peripheral nerve sheath tumours (MPNST), which displays rhabdomyosarcomatous differentiation. It was first described by Masson in 1938. The MTT term was introduced by Woodruff in 1973 based on the experimental work of Locatelli who noticed a growth of supernumerary limb when he transplanted the sciatic nerve on the dorsal surface of triton salamander. MTT commonly originates in the nerves of extremities, the trunk, head and neck, less commonly in the mediastinum and very rarely in intercostal nerves.²

CASE PRESENTATION

A man aged 28 years, a non-smoker, presented with pain in the right hypochondrium gradually worsening over the last 5 months. There was no history of vomiting, cough, shortness of breath, loss of weight or appetite. On physical examination, he had café-au-lait spots on his trunk, and palpable small subcutaneous swellings along the peripheral nerves of both arms. Chest examination revealed decreased air entry on the right lower chest and a palpable non-tender mass extending into the right hypochondrium. Ultrasound scan of the abdomen

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Figure 1 (A) Preoperative CT scan thorax revealed a large mass originating from the chest wall and compressing the liver. (B) Postoperative CT scan thorax showing no recurrence and liver regained its normal anatomical position. (C) Gross specimen 15×8 cm showing intercostal nerve sheath tumour en bloc with ribs, part of diaphragm and peritoneum. (D) Chest wall reconstructed.

New Jersey, USA) at all bony margins. The margins of MMS were sutured with continuous 2/0 prolene to the surrounding tissues (figure 1D). Chest wall wound was closed by approximating the soft tissues; skin was closed without using any myocutaneous flap. As the diaphragm defect was peripheral measuring 8×5 cm, there was no need to reconstruct it with any prosthetic material. This is a very useful technique to restore chest wall stability and diaphragm integrity and function, which had not been described earlier. The patient was extubated in the operating theatre.

OUTCOME AND FOLLOW-UP

His postoperative recovery was uneventful and was discharged home on the eighth postoperative day. Gross pathological examination showed a rounded mass measuring 15×12×8 cm (figure 1C). Histology report showed spindle mesenchymal cell proliferation with increased mitotic activity (figure 3A). There were foci of rhabdomyoblastic differentiation (figure 3B). Immunohistochemical stains show these cells to be positive for desmin and myogin (figure 3C). His follow-up CT scan of the chest after 1 year revealed no recurrence of tumour and the liver had regained its original anatomical position (figure 1B).

DISCUSSION

MPNST accounts for 5% of all soft tissue sarcomas and 30–50% of these are associated with NF1. The incidence of MPNST is 4.6% in patients with NF1 and 0.001% in patients without NF1.³

MTT is a rare divergent subset, which accounts for <10% of MPNST, commonly occurs in young people and 50–70% of these patients have NF1, although this can occur sporadically in 25–30% of cases without NF1 disease predominantly in old age women.⁴ Clinical findings are usually not significant until they rapidly enlarge to produce pressure effects or they invade the neighbouring structures.

CT and positron emission tomography scans are the imaging modalities used for initial diagnoses and post-treatment surveillance. Final diagnosis is based on histopathological and immuno-histochemical staining report of gross specimen because sometimes the fine needle aspiration or true cut biopsy cannot provide the precise diagnosis. The best treatment option is radical excision aiming for tumour-free margins. Incomplete and cytoreductive resection in patients with MTT is not recommended neither has shown any survival benefit. Radical surgical resection with negative margins is the single most defining factor for long-term prognosis. MTT is biologically very

Figure 2 Illustration of improvised surgical technique.

Figure 3 (A) Malignant peripheral nerve sheath tumour with proliferating malignant spindly tumour cells (H&E $\times 200$). (B) An area of rhabdomyoblastic differentiation. Note rounded rhabdomyoblasts with eosinophilic cytoplasm. (H&E $\times 200$). (C) Myogenin stain featuring nuclear stain of rhabdomyoblasts (magnification $\times 200$).

aggressive in nature, has a high propensity for early local recurrence and distant metastasis after resection. These tumours have extremely poor prognosis and overall survival is 33% and 12% at 2 and 5 years, respectively.⁵ Angelo *et al*⁶ reported better prognosis with 5 cm or more tumour-free resection margin. Positive resection margin is a worse prognostic factor independent of tumour size and site and age of the patient.⁷ Surgery is the mainstay of treatment, the role of radiotherapy or chemotherapy is not well established, although a less local recurrence and better survival has been reported in patients who received adjuvant radiotherapy.⁸ After extensive chest wall resection, a geometrical and stable reconstruction without compromising the respiratory physiology is a challenging task. Methyl methacrylate cement sandwiched in a Marlex mesh moulded accord-

ing to the geometry of the chest wall is one of the best materials to provide optimal reconstruction with maximum stability. The most important drawback of this material is tilting and excursion of the plate if it is not properly fixed.

In conclusion, MTT of intercostal nerve sheath in a patient with NF1 is a very rare entity. The mainstay of the treatment is surgery to achieve a curative resection by radical en block excision of the ribs and involved structures. Our improvised surgical technique is an excellent method to restore aesthetic contour and stability of the chest wall and diaphragm integrity with a minimal risk of plate excursion and dislodgement. Primarily diaphragm integrity was restored without the use of any synthetic material which is very useful method in such cases.

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Contributors IUC, operating surgeon, performed the procedure and wrote the abstract and manuscript. AIC is the assistant surgeon and searched for the references. HM wrote the part of the discussion and helped in cropping the pictures. SSA, pathologist, provided the histology report and slides.

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Patient's perspective

The patient was happy with the surgical outcome and had a better quality of life. He refused adjuvant chemotherapy and radiation therapy.

Learning points

- ▶ Triton tumour of intercostal nerves is a highly malignant neoplasm; its diagnosis needs specific immunostaining.
- ▶ Radical surgical resection with wide tumour-free margins is the mainstay of treatment.
- ▶ These tumours are resistant to chemotherapy and radiation therapy.
- ▶ Continuous surveillance with follow-up CT scans is essential as there is high propensity of local and distant metastasis.
- ▶ Our improvised reconstruction technique is an excellent method to restore chest wall stability and diaphragm integrity with minimal risk of MMS plate excursion and dislodgement.

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