Clinical Records

Neurofibrosarcoma

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NEUROFIBROSARCOMA as a distinct entity, apart from being a common malignant tumour, has been recognized for a long time. In 1936 Foot described the pathological changes and there were earlier papers by Stewart and Copeland (1931), and by Stout (1935).

The site of origin of the tumour is variable and in the series reported by Hosoi (1931) there were two in the neck and one each in the thigh, the retroperitoneal structures, the shoulder, the sacrum and the eyeball. In many patients with numerous cutaneous neurofibromas, no sarcomata were present. On this evidence there seems to be no definite relationship between sarcoma and the number of cutaneous lesions.

The tumour arises from the Schwannian cells of neurolemma. It may occur at any age but is commonest after the fifth decade of life. It has a definite and specific association with multiple neurofibromatosis. It is estimated that about half of the malignant neurofibrosarcomas arise in persons with disseminated neurofibromatosis. It not only diffuses into and infiltrates the surrounding tissues but metastasizes readily through the blood stream to the chest, bone, liver and brain. Lymphatic invasion is uncommon.

The tumour may form a fusiform dilatation of a nerve trunk and destroy the nerve inside the sheath. Since it is not limited by a capsule, there is insidious and persistent infiltration into the surrounding tissues. The tumours occurring in Von Recklinghausen's disease frequently undergo sarcomatous changes (50 per cent.). Penfield and Young (1930) stated that, clinically, neurinoma appears as a tumour with a decided tendency to undergo sarcomatous degeneration which is further evidence of the mesodermal character of the original tumour.

Treatment of patients with sarcoma in most cases is unsatisfactory. Some patients with sarcoma apparently are cured by surgical removal of the tumour as is shown in the series of Preston *et al.* (1952). Recurrences are seen developing in cases after operation. It is evident that most of these patients reported for treatment only after the sarcomas had become large and had been present for many months.

Case History

A male patient aged 22 years was admitted on 2 June 1966 with the complaints of hoarseness of voice and gradually increasing swelling on right side of neck, the duration of which was $2\frac{1}{2}$ months.

There was no history of cough with expectoration, loss of weight, loss of appetite, haemoptysis or low grade fever.

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On examination there was large swelling on the right side, $5 \times 2\frac{1}{2}$ in. in size, oval in shape with a nodular feel and firm in consistency (Figs. 1, 2 and 3). The growth was fixed to the underlying structures. There was no ulceration, no signs of inflammation, no sinuses and no scar tissue. Indirect laryngoscopy showed that he had a complete right vocal cord paralysis.

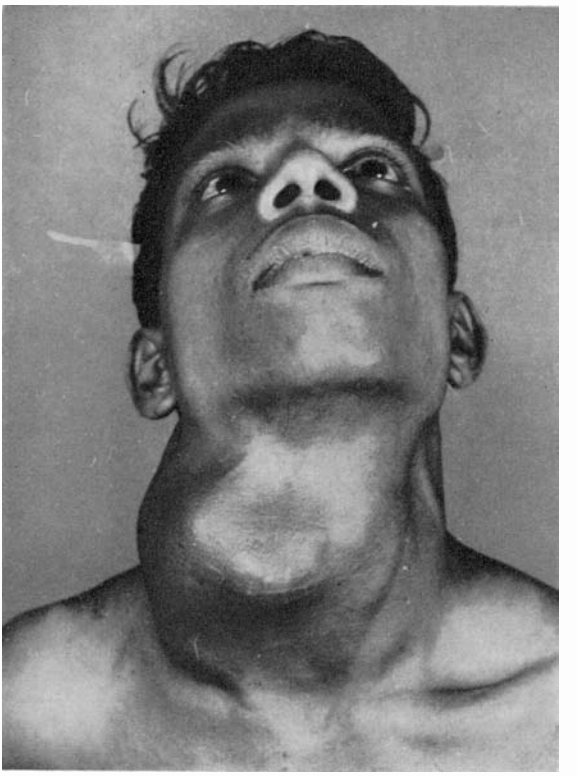


FIG. 1.
Patient with swelling.

On systemic examination there was no evidence of ascites or oedema, but there were small swellings all over the body with pigmented patches typical of Von Recklinghausen's disease, Routine blood count, blood sugar and E.C.G. investigations were within normal limits. X-ray revealed an apical shadow on the right side. The shadow was in continuity with tumour shadow in the neck (Fig. 4).

In the presence of multiple neurofibromata, a tentative diagnosis of a sarcomatous change in a neurofibroma was made. Biopsy confirmed the diagnosis of neurofibrosarcoma (Fig. 5).

Considering the fixity of the tumour it was not a good case for any operative procedure. Having had good results with chemotherapy in early fixed metastasis

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FIG. 3.
Tumour and neurofibroma on the chest wall below the left nipple along the seventh rib. Pigmented area along the left ninth rib.

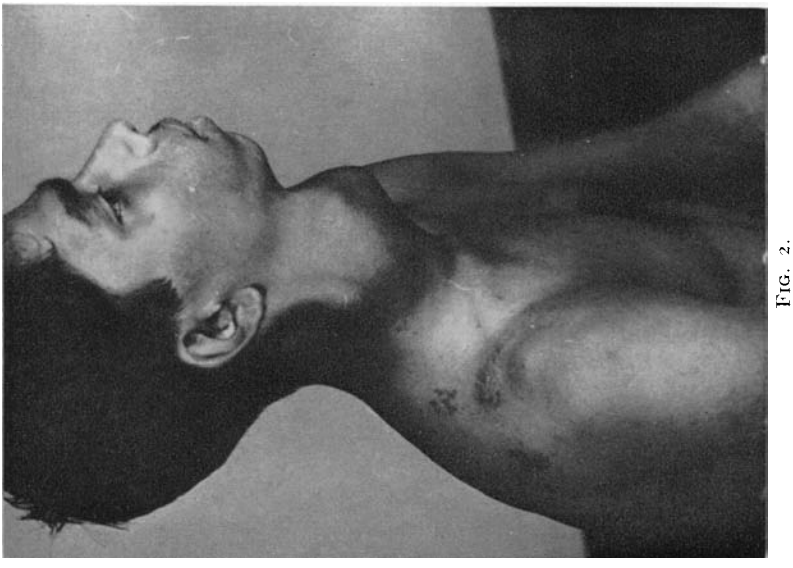


FIG. 2.
Patient with swelling.

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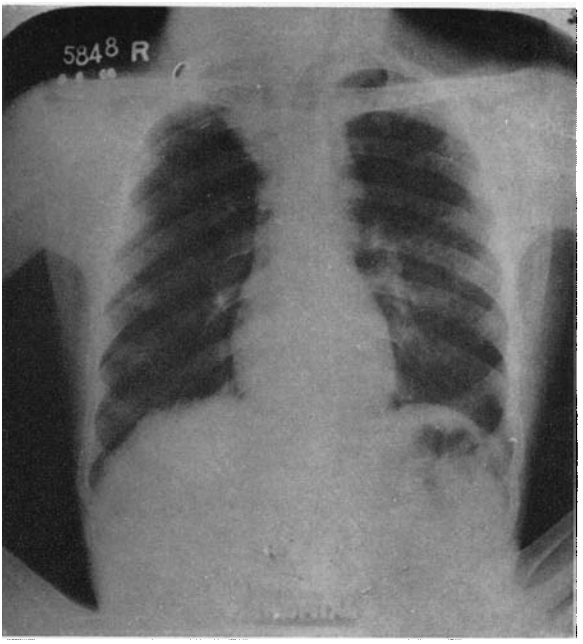


FIG. 4.
X-ray shows apical shadow on the right side. The shadow was in continuity with tumour shadow in the neck.

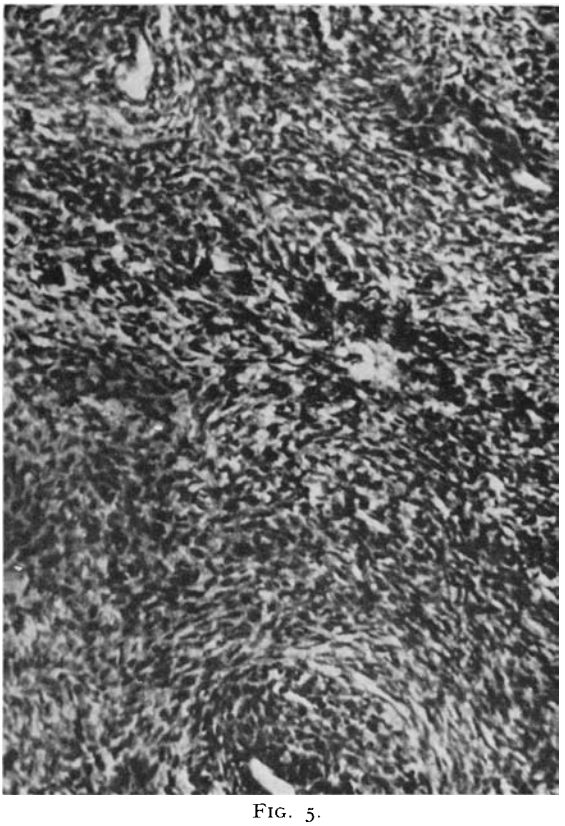


FIG. 5.
Biopsy report: Section consists of a very cellular tumour which was formed in whorls. In places there is a suggestion of regimentation of nuclei which are hyperchromatic and plump. There are one or two areas of necrosis.

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(Hiranandani, 1966), it was decided to try chemotherapy and see if it made any difference in the present case. After a full dose of Endoxan, there was no change in the tumour. The patient's condition was already bad and it was decided to operate, even though it meant that it would mean sacrificing the internal carotid artery which was expected to be already compressed by the tumour.

Operation

Under endotracheal anaesthesia the operation was performed. Four pints of blood were required. At first the upper part of tumour was exposed. The internal carotid artery above the tumour was dissected out with the vagus nerve (Figs. 6a and b). The lower part of tumour was then dissected (Fig. 7). It was adherent to the laryngeal framework from which it was easily dissected. Internal carotid artery was completely engulfed and compressed by the tumour but was not totally obliterated. The only solution of the problem was to ligate the artery

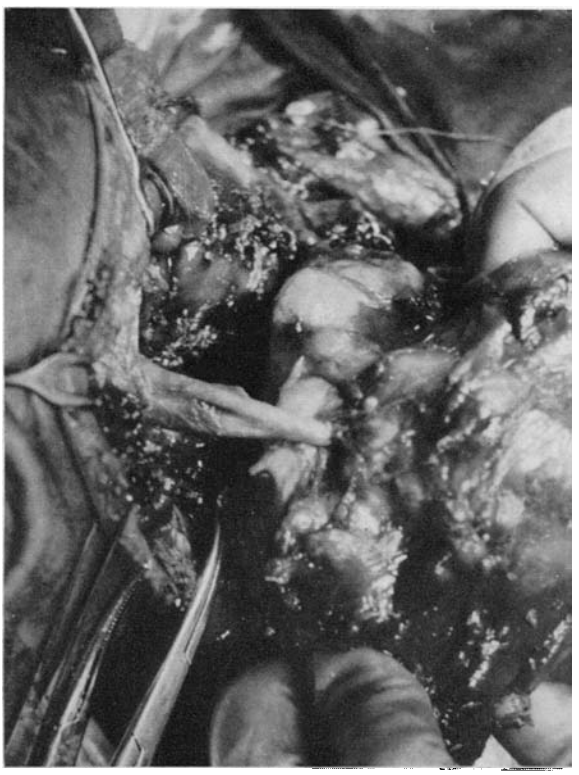


FIG. 6a.

Internal carotid artery above the tumour and its two terminal branches dissected out.

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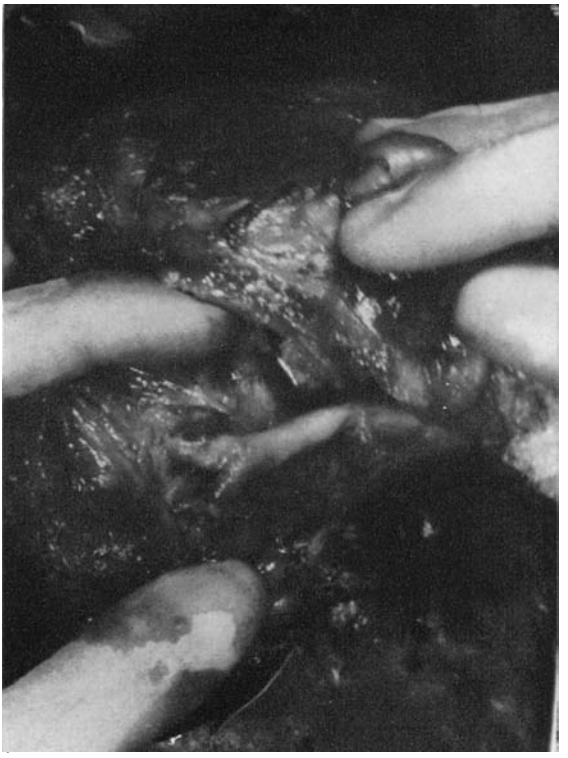


FIG. 6b.
The tumour dissected out and pushed medially to expose the proximal part of artery.

below and above the tumour which was done. The subclavian artery was also passing through the tumour and was therefore ligated proximal and distal to it (Fig. 8). The tumour then was seen going into the thoracic cavity behind the sternoclavicular joint. While trying to separate and remove the tumour from the chest cavity, the left subclavian vein was ruptured but it was carefully sutured. The apical pleura was ruptured (Figs. 9 and 10). A drain was put in the chest cavity because of tear in the pleura and subsequent bleeding in the chest cavity. The tumour was then removed (Fig. 11). Only at one place of the thoracic extension was a slight raw area seen (Figs. 12, 13 and 14). Since the whole tumour from the neck was completely removed one expected this patient to have a good chance but one was not happy with the lower portion of the tumour because it was possible that a small portion of it might have been left at the chest cavity. The patient made a complete uncomplicated recovery. He did not develop any hemiplegia. Unfortunately, on the fifteenth day he developed a sudden haemorrhage from the supraclavicular region. The region was explored but the bleeding had already stopped and a few gelfoam pieces and gauze packing were placed over the site of bleeding. Four days after that he had another bout of bleeding and expired.

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FIG. 8.

The proximal part of common carotid and subclavian and distal part of common carotid arteries being clamped.

FIG. 7.

The tumour is dissected medially from the floor of posterior triangle and from the laryngeal framework. It shows the proximal part of common carotid artery dissected out, passing through the tumour and the artery emerging above the tumour.

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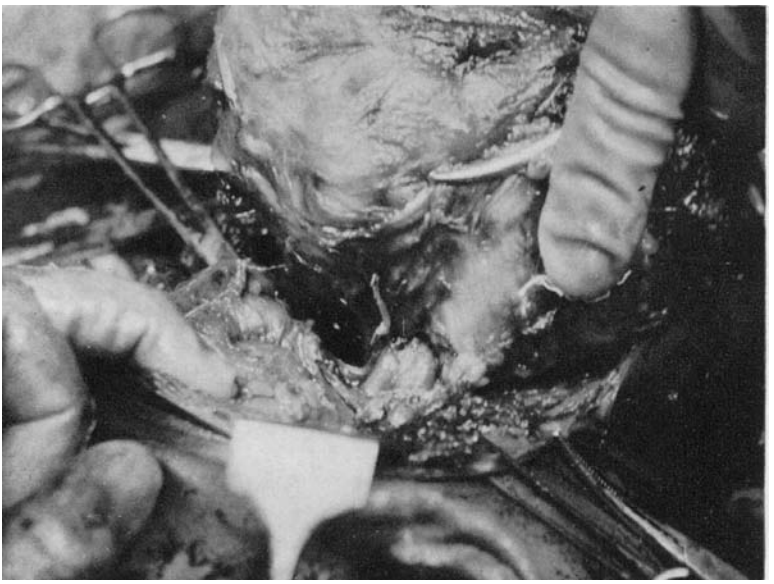


FIG. 9.

Shows an oval opening into the pleura from which the tumour has been dissected out.



FIG. 10.

The lower part of tumour, dissected out from the chest wall, shows an ulcerated area.

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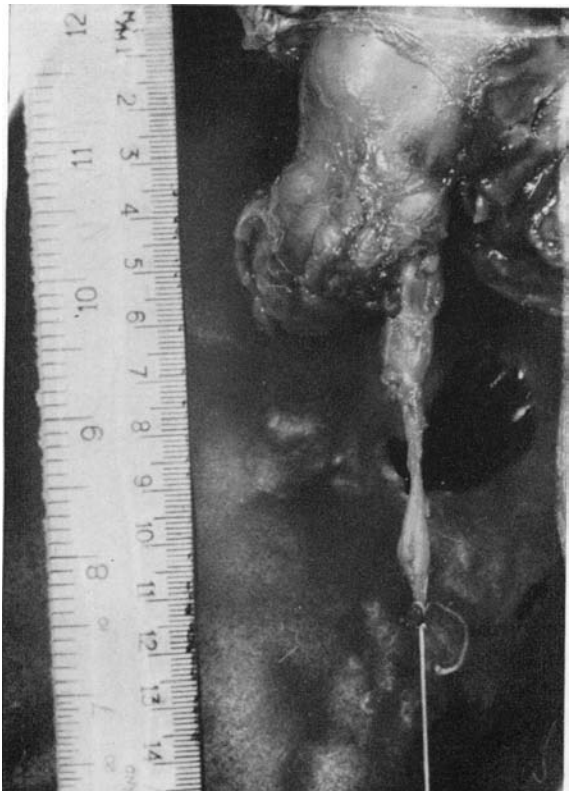


FIG. 11.
Lower part of the tumour removed from the chest with the vagus nerve, showing infiltration of its proximal part and the opening in the apical pleura.

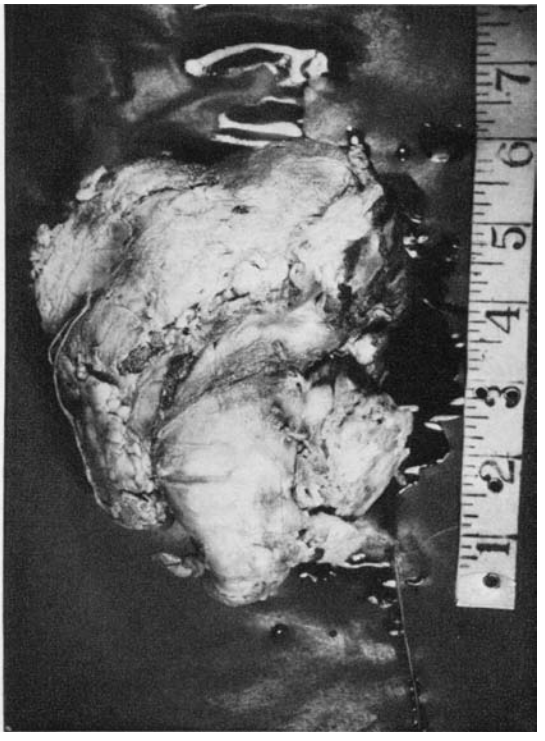


FIG. 12.
The tumour with its external surface.

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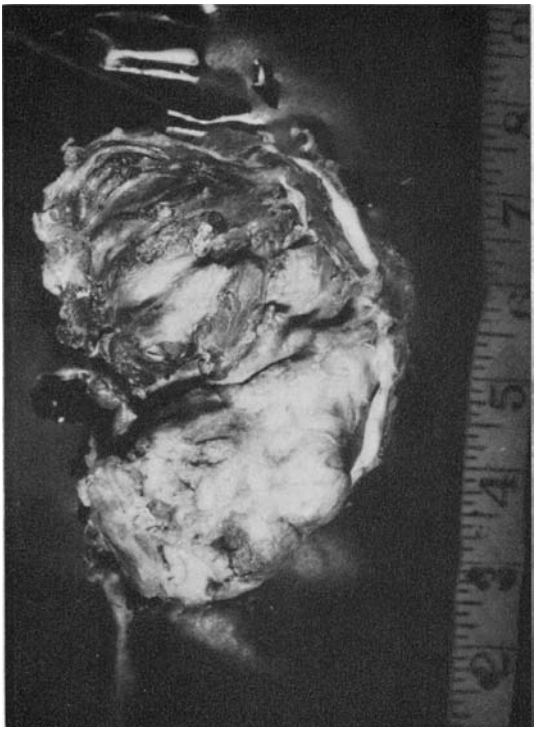


FIG. 13.
Tumour showing the internal surface.



FIG. 14.
Cut section of the tumour showing the main mass, the compressed common carotid artery and the proximal infiltration of vagus nerve.

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Post-mortem findings

An ulcerated growth $1 \times \frac{3}{4}$ in. was seen in the posterior part of chest wall in the apical region (Figs. 15, 16 and 17). This was the place from where the original growth was torn and removed. The bleeding bout was from the growth itself. The cause of death was post-operative bleeding from the ulcerated mass.

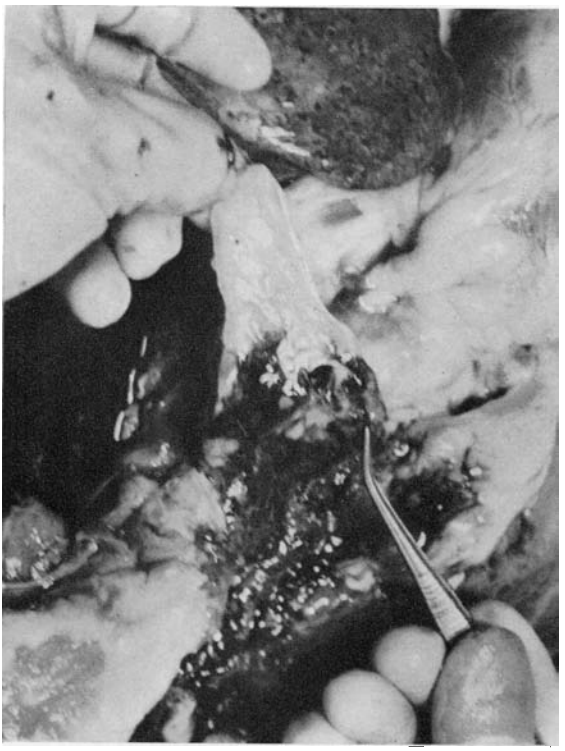


FIG. 15.
Posterior part shows ulcerated growth.

Discussion

In 1930 Lewis and Hart described four cases and opined that sarcomas are extremely malignant and tend to form metastases along the nerve primarily involved. They may extend to other nerves passing to these, apparently along branches, communicating with the nerves primarily involved. They described in a 19-year-old male patient neurofibrosarcoma of right vagus filling the space between angle of jaw and the mastoid occupying the upper half of neck anteriorly. Patient died 16 months after operation.

Hosoi (1931) reviewed sixty-five cases of sarcoma in Von Recklinghausen's disease, reported in the literature. Borberg found only one patient with sarcoma

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in seventy-six living patients with the diseases whom he examined. In the series of Preston, Walsh and Clarke in 1952, ten persons developed the sarcomatous change in Von Recklinghausen's disease. Out of these only one was found in the neck region.



FIG. 16.
Ulcerated growth with a big vessel from which patient had bled to death.

This case reported here according to his history, developed a rapidly enlarging tumour within 2½ months. The tumour formed a separate nodule along the nerve. Though it started in the neck and attained a large size, except for slight adhesions to the laryngeal framework, it could be easily dissected out in the neck. However, when it extended into the chest cavity it probably got ulcerated due to decreased blood supply distally and the ulcerated mass became secondarily adherent to chest wall. In the apical region, the tumour showed a constricting ring which must have also contributed to the decrease in blood supply.

According to the available literature, in 1930 Lewis and Hart described a similar tumour in a patient who died 16 months after operation. If the patient was seen earlier before the tumour extended into the chest cavity, the authors feel it would have been easy to excise it completely and he would have had a

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good chance of survival. Because, even at such a late stage, very careful autopsy did not reveal any lymph or blood borne metastasis.

Summary

A case of neurofibrosarcoma is reported. This is the second young patient described in the literature so far. A careful autopsy did not reveal any blood borne or lymph metastases.

Acknowledgments

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