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VERTEBRAL ANGIOSARCOMA

A Case Report

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Angiosarcoma (syn. malignant hemangioendothelioma) of bone, especially of the vertebrae, is a rare neoplasm. Bundens *et al.* (1965) reviewed 32 well-documented cases of bone angiosarcoma, out of which only 3 appeared in the spine. Ackerman *et al.* (1962), De Rubertis *et al.* (1967), and Krueger *et al.* (1961) reported 4 additional cases.

Since the tumour is rare, it might be justified to report one more case of vertebral angiosarcoma to demonstrate the dramatic course of the disease.

CASE REPORTS

A 19-year-old female typist was admitted to the Department of Orthopaedics and Traumatology of the University Central Hospital, Helsinki, for examination. For over a month she had experienced pain between the scapulae and in the shoulders; for two days before admission she had found it difficult to walk, and the pain had expanded to the lumbar area. She was unable to defecate and micturate. The day previous to admission she was bedfast, because of immobility of the right leg. She complained of severe throbbing pain in the back at the level of the lumbar spine and on both sides of the chest when any effort to move was made. She experienced a tingling sensation in the chest wall and right hand, and was unable to laugh or cough.

On arrival at the hospital the right leg was completely flaccid, the patient was unable to lift the leg or to move the toes or the foot. The Lasegue sign was positive at 75°. The patellar and achilles tendon reflexes of the right leg were active, no clonus. The Babinski sign was negative. The entire area of the right lower extremity and the right side of the abdomen were hypersensitive. The left leg had normal strength. The Babinski sign was negative. The other reflexes were medium active, the Lasegue sign was positive at 75°. The pulses and the skin temperatures of the legs were normal. In the area of T11 on the back an extremely tender tumour, about 6-7 cm in diameter was palpable.

About 12 hours after admission a total paralysis had developed below the level of the 4th rib and the reflexes were completely lacking. There was an anaesthetic

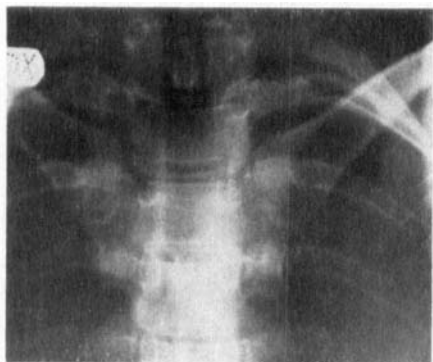


Figure 1



Figure 2

*Figure 1. Roentgenogram. Destruction of spinous process in TII.
Figure 2. Tomogram. Cyst-like formation on the right side in vertebral body of TII.*

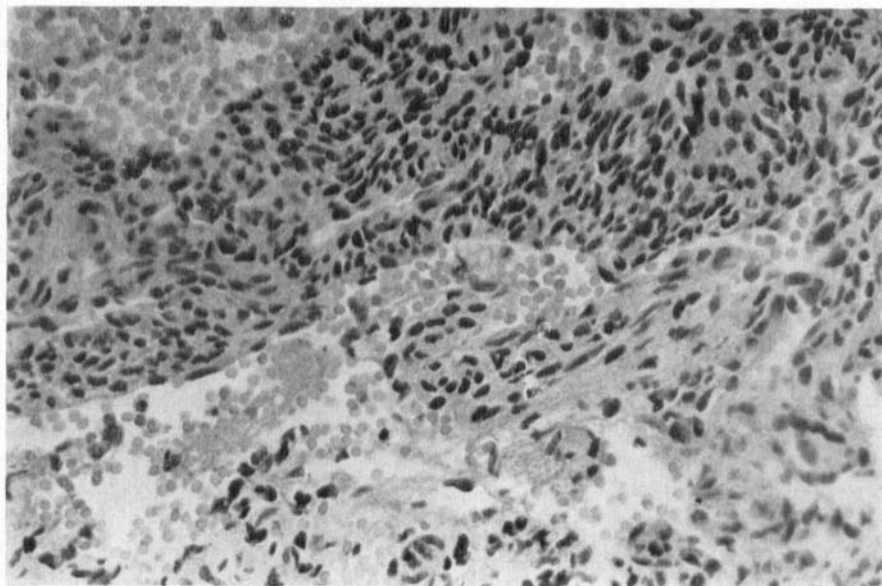


Figure 3. Microphotograph. Anastomosing vessels, and abundant atypical cells of endothelial origin. Magnification $\times 250$. Weigert—Van Gieson staining.

area extending caudally from the 4th rib over all segments. X-ray examination (Figure 1) revealed a destruction of the lamina and a partial destruction of the spinous process of TII. The tomogram (Figure 2) showed the presence of a round opaque area, about 2 cm in diameter, in the vertebral body, and a destruction of the right half of the spinal arch and spinous process of TII. Since paralysis had developed rapidly and was complete, an immediate operation was performed.

Longitudinal incision through the layers of tissue over the area of T1-T11 revealed a fluctuating tumour, about 7 cm in diameter, which was partly formed of a necrotic mass and blood. After removal of the tumour, tissue engorged with blood was found in the bottom of the cavity. The tumour was located in a region corresponding to the right spinal arch and spinous process of T11, which were completely destroyed. The left spinal arch had also been destroyed to some extent. Similar changes were present in the right pedicle and the vertebral body. The bone was bleeding profusely and could only be staunched with bone wax with difficulty. The tumour was so extensive that radical extirpation was impossible. The exposed dura mater in the bottom of the cavity was apparently intact, although compressed by haematoma. Biopsies were taken from the tumour. A suction drainage was placed and the wound closed. The neurological state remained unchanged after the operation. The patient died suddenly on the 4th postoperative day. The autopsy revealed a primary malignant tumour of vascular origin which had destroyed its surroundings. Extradural haematoma had extended to the cervical spine and had compressed the medulla all the way to the respiratory centre. It was concluded that the patient died from a respiratory arrest.

The pathological diagnosis was angiosarcoma (Figure 3). The characteristic features were abundant anastomosing blood-filled tubes lined with proliferating atypical cells of endothelial origin, which infiltrated the surroundings, including the bone. At some places the neoplastic tissue revealed sarcomatous make-up and papillary formations.

DISCUSSION

In his paper on angiosarcoma Stout (1943) describes the condition as a neoplasm made up of congeries of vascular tubes which have a marked tendency to anastomose and are lined with hyperchromatic tumour cells of endothelial origin. Jaffe (1958) and Leichtenstein (1965) agree that angiosarcoma is practically impossible to diagnose clinically, i.e., without an operation and subsequent histologic examination of biopsies. The case presented here shows features indicating, according to the above authors, that the tumour is an angiosarcoma. This case closely resembles that reported by Krueger *et al.* (1961). In both instances the patients were young adults who revealed similar subjective symptoms and clinical signs. The location of the tumour was almost the same in both cases.

As for the treatment of angiosarcoma when diagnosed, most authors (Bundens *et al.* 1965, Carter *et al.* 1956, Jaffe 1958, Krueger *et al.* 1961, Lichtenstein 1965) recommend primarily radical surgery whenever possible, combined with massive postoperative X-ray irradiation. Morgenstern (1960), on the other hand, reports a case of angiosarcoma of the clavicle cured with irradiation only, and recommends radiation as the sole treatment.

The prognosis of angiosarcoma is poor for several reasons. Out of

the 32 cases reviewed by Bundens *et al.* (1965), only 9 were known to have survived until the time of reporting the cases; however, none had an angiosarcoma of the vertebrae. There is only one case of vertebral angiosarcoma in the literature (Krueger *et al.* 1961) where the patient was known to be alive 2 years postoperatively. Since the disease in the present case was already so far advanced by the time of admission to the hospital that a beneficial radical operation was impossible, a decompressive measure was taken. Because the case quickly terminated fatally, no irradiation treatment could be started.

SUMMARY

A case of vertebral angiosarcoma (malignant hemangioendothelioma) is presented. Only 7 cases of this type have been reported earlier. The case involves a 19-year-old female with a histologically verified angiosarcoma of vertebra TII. The compression of the medulla caused by profuse extradural bleeding of the tumour was lethal.

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