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PRIMARY EXTRAOSSEOUS OSTEOGENIC SARCOMA IN THE RETROPERITONEAL SPACE

A case report and review of the literature

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Abstract

One case of primary, extraosseous osteogenic sarcoma in the retroperitoneal region is presented, which is the tenth case hitherto reported in the literature. The 3 females and 7 males reported had a median age at diagnosis of 59½ years. The tumours were all large at diagnosis and the symptoms short and uncharacteristic. There was no characteristic set of laboratory or roentgenologic findings; 2 of the 10 cases had elevation of alkaline phosphatase, and in 4 cases roentgen examination revealed tumour calcifications. The prognosis was poor and 8 of the 10 patients died, with a median survival of 9½ months. In 3 cases, surgery was supplemented by chemotherapy without obvious improvement in survival.

Key words: Soft tissues, neoplasms, retroperitoneal space, extraosseous osteosarcoma.

Extraosseous osteosarcomas are rare, and only about 120 cases have been described in the medical literature; about 120 other cases are registered in the archives of the Armed Forces Institute of Pathology (7).

Very rarely, an extraosseous osteosarcoma is located in the retroperitoneal region, and only 9 cases with this location have so far been reported (1, 2, 5, 8, 10, 12, 14, 19). A further case is described in the present report and a review of the literature is also presented.

Case report

A 56-year-old, previously healthy man was admitted to the Central Hospital in Herning. He had felt tired for 4 months and during the past 6 weeks had suffered from progressive pain arising below the curvature of the rib on the right side, where he had a feeling of distention. A large, firm mass was palpable in the right upper part of the abdomen just below the rib curvature. The sedimentation rate was 42 mm/h, and the alkaline phosphatase 357 U/l (normal range 50–275 U/l). Serum calcium, liver enzymes, and routine urinary tests were normal.

Abdominal radiography showed a 25 cm × 16 cm soft tissue mass without calcifications, located at the site of the right kidney. Ultrasonography revealed a collection of well-demarcated, echovoid, confluent cavities. The kidney was poorly defined and appeared larger than the contralateral one; the lower renal lobe was seen 1 to 2 cm below the umbilical level. Urography showed compression of the right renal pelvis and dispersion of the renal calices over the large soft tissue mass. The left kidney appeared to be normal. Chest radiography showed a rounded density in the right inferior lobe, approximately 25 mm in diameter.

The tumour, the right kidney, and the proximal part of the right ureter were excised. The tumour, well-demarcated without bone involvement, weighed 4.5 kg. No signs of intra-abdominal tumour spreading were observed during the operation.

Gross pathology. The renal parenchyma, pelvis, and ureter appeared normal. The renal surface at the upper renal pole was compressed medially by a rounded, non-encapsulated but apparently well-demarcated tumour, 20 cm in diameter. The renal parenchyma did not seem to be infiltrated. Section through the tumour showed a large, irregular, partly clot-filled cavity, encircled by grey-white tumour tissue with a consistency varying from soft to firm, with small firm areas of 3 to 10 mm. The tumour tissue contained both small and large necrotic areas (Fig. 1).

Histopathology. The tumour extended into the renal capsule, but did not penetrate it. The ureter was partly surrounded by the tumour but was not infiltrated. The large, central parts of the tumour were partly or completely necrotic, while the peripheral tumour tissue was better preserved. Samples were taken from several parts of the tumour and in all sections it had the character of an osteosarcoma with varying degrees of differentiation. Some areas contained well-formed osteoid trabeculae separated by hypercellular stroma composed of moderately polymorphic and sparsely mitotic spindle cells. Other areas contained very small, dispersed, and irregular trabeculae encircled by spindle cells and with up to 10 mitoses/10 HPF. Multinucleated cells were sporad-

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ic, notably seen near osseous trabeculae, and chondroid elements were scarce (Fig. 2).

Radiography of the entire skeleton revealed no tumours. Forty-five days after excision of the retroperitoneal tumour parts of the middle and lower lobes of the right lung containing osteosarcomatous metastases were resected. Fourteen days later, fat and connective tissue under the right dome of the diaphragm was removed; it also contained metastases of osteosarcoma.

Three months after the first operation, exploratory laparotomy revealed local tumour recurrence and a chain of lymph node metastases along the aorta. All visible tumour tissue was resected.

Radiation therapy and postoperative adjuvant chemotherapy was not administered owing to the disseminated nature of the disease.

The patient died at home three months after the last operation. Autopsy was not performed.

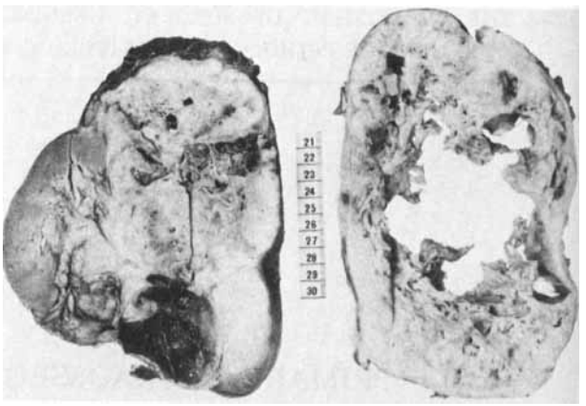

Fig. 1. Sections through the peripheral part of the tumour showing its relation to the right kidney (left) and through the middle part of the tumour showing central, necrotic cavity (right).

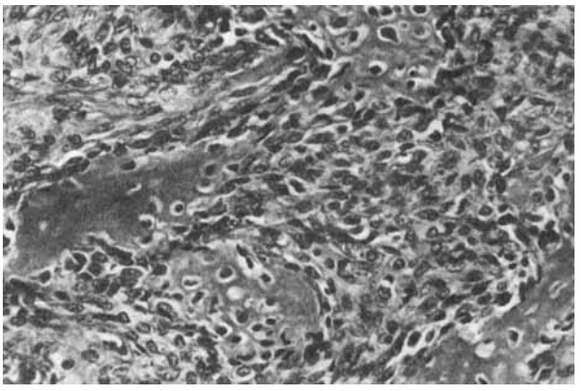

Fig. 2. Section of tumour showing osteosarcoma composed of spindle-formed cells and irregular, osteoid trabeculae. H & E ×125.

Discussion

The diagnosis of primary, extraosseous osteogenic sarcoma (PEOS) rests on three criteria (18): 1) the presence of a morphologically uniform sarcomatous tissue which eliminates the possibility of a mixed malignant mesenchymal tumour; 2) sarcomatous tissue, producing malignant osteoid; 3) the ruling out of any osseous origin. The present case seems to meet all three criteria. Concerning the first criterion it might be difficult to exclude a malignant teratoma with one-sided development into osteogenic sarcoma. However, samples were taken from several parts of the tumour and all sections showed the picture of osteosarcoma. Radiography of the whole skeleton did not reveal any bone tumours. Autopsy would have been of value but could unfortunately not be performed.

Primary soft tissue osteosarcomas account for a mere 1.2 per cent of all soft tissue sarcomas (1) and 4.6 per cent of all osteosarcomas (15). In 100 patients with PEOS collected from five series (1, 6, 15, 17, 20) the dominant sites were the thigh (46%), the upper extremities (12%), and the nates (11%). With the present case included, the number of retroperitoneal PEOS described in the medical literature totals 10 cases (Table).

Possible factors predisposing to PEOS include previous radiation therapy (3, 4), myositis ossificans (8, 10, 13, 16), traumas (1), thorotrast injection (9), and intramuscular penicillin injection (11). However, in most of the reported cases these potentially predisposing events were lacking. In 3 of the 10 reported cases with retroperitoneal PEOS potentially predisposing factors were found. Patient No. 3 had a history of radium treatment and hysterectomy for endometrial carcinoma 7 years and cholecystectomy 5 years before diagnosis of PEOS. In patient No. 6 a 2 cm calcification was seen at the later tumour site; it possibly represented myositis ossificans. Patient No. 9 underwent hemicolectomy for adenocarcinoma coli 3 years before diagnosis.

The median age of the 10 patients with retroperitoneal PEOS was 59½ years. The median age for patients with soft tissue osteosarcomas as a whole was found to be 51 years in a series of 48 patients (17) and 53 years in four series with a total of 52 patients (1, 6, 15, 20). This age disparity between retroperitoneal PEOS in general is not statistically significant. The group of patients with retroperitoneal PEOS comprises 3 women and 7 men, a sex ratio (M:F ratio) that does not significantly differ from the M:F ratio of 0.89 for the PEOS group as a whole.

The symptoms reported in retroperitoneal PEOS were, as in other retroperitoneal tumours, uncharacteristic. The duration of symptoms was short, with an average of about 2 months. The tumours were often large at diagnosis. Radiography demonstrated tumour calcifications in 4 of the 10 cases (patients Nos 3, 5, 6, and 9). Radiographic examination is reported to show calcifications in approximately half of all soft tissue osteosarcomas (15). Two patients (Nos 9 and 10) with retroperitoneal osteosarcoma had preoperative elevation of alkaline phosphatase, an elevation that could not be ascribed to any other disease.

Table

Age and sex, duration of symptoms, possible predisposing factors, tumour size, treatment, and survival in 10 patients with retroperitoneal PEOS

Case no.	Ref. no.	Sex and age	Symptoms (duration)	Possible predisposing factor (years before)	Tumour diameter (cm)	Treatment	Survival (follow-up time after diagnosis)
1	8	M 58	–	–	–	Excision	Alive and well (36 months)
2	2	M 53	–	–	–	Excision, irradiation, chemotherapy	Dead (36 months)
3	5	F 65	Nausea, vomiting (2 months)	Radium implant into uterine cavity and hysterectomy (7) cholecystectomy (5)	14	Excision	Dead (14 days)
4	12	M 68	Pain in back and right leg	–	6	Excision, irradiation, chemotherapy	Dead (11 months)
5	14	M 61	Weight loss (3 months)	–	12	Excision	Dead (4 months)
6	10	F 55	Abdominal pain (3 months)	Myositis ossificans (5)	20	Excision	Dead (10 months)
7	1	F 63	Swelling and pain (2 months)	–	–	Excision, irradiation	Dead (27 months)
8	1	M 41	Swelling and pain (2 months)	–	–	Excision, irradiation, chemotherapy	Dead (9 months)
9	19	M 85	Abdominal pain (24 hours)	Adenocarcinoma coli with hemicolectomy (3) cholecystectomy (3)	35	Excision	Alive and well (8 months)
10	Present series	M 57	Fatigue (4 months), abdominal pain (1½ months)	–	20	Excision	Dead (8 months)

Alkaline phosphatase was normal in one case and in the remaining 7 cases no values were mentioned. For soft tissue osteosarcomas in general, preoperative elevation of alkaline phosphatase in the range 22 per cent (15) to 83 per cent (13) has been reported.

Excision was performed in all 10 cases. In 3 cases, excision was supplemented by chemotherapy and irradiation, and in one case by irradiation alone; the malignant disease proved fatal in all of these 4 patients, median survival being 19 months. The prognosis was on the whole poor, and the disease was fatal in 8 of the 10 patients, with a median survival of 9½ months. Patients Nos 1 and 9 were alive without symptoms 36 and 8 months, respectively, after surgical resection.

Among the 45 patients with PEOS in general, followed for more than one year, 41 had local recurrence (69%) and/or distant metastases (80%), and the median survival was 31 months (17). It is not known whether more aggressive multidisciplinary treatment of the type often used for bone osteosarcoma (multi-drug chemotherapy, radiation therapy, surgery) has anything to offer in PEOS.

Due to its extreme rarity, retroperitoneal PEOS is, as a rule, not suggested on purely clinical and roentgenologic grounds. The possibility of this tumour might, however, be considered notably if a retroperitoneal process shows roentgenologic calcifications and/or if the serum alkaline phosphatase is elevated without signs of liver or bone involvement; nevertheless, negative findings will not rule out the diagnosis. From the histopathologic point of view the most important differential diagnoses are: 1) other sarcomas with calcification and osseous metaplasia; 2) mixed tumours (teratomas) with osteosarcomatous parts; and 3) myositis ossificans with zonal presentation, more or less marked central polymorphy, and well-differentiated, peripheral ossification.

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