ANGIOLEIOMYOSARCOMA – A VERY RARE MALIGNANT TUMOUR FROM THE SOFT TISSUE

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ABSTRACT

In the article we describe a case of a 12- year old boy with the tumour in the small pelvis. The only one abnormality in laboratory tests was a 3- numeral ESR and a big solid tumour probably extended from the urine bowel, found on radiological examination. On surgery the tumour was found to extend from the ascendent colon. On pathological examination, it was described as angioleiomyosarcoma with metastases in the lymph nodes. The boy was treated with IX courses consisting of ifosfamide, vepeside or ifosfamide, adriamycine and vincristine. At present, the child is in the first complete remission, 7 months after completion chemotherapy.

INTRODUCTION

Angioleiomyosarcoma is a very rare malignant tumour originating from the smooth muscular and angiopoetic tissues. It occures mainly in medium and old persons. It constitutes about 1% in the soft tissue tumours. The neoplasm is unusual in the childhood. The prognosis, regardless of localization and age, is very poor. Case: 12-year old boy was admitted to our hospital because of 12-months lasting pallor and weakness with palpable, hard, unfixed tumour in the small pelvis. The diameter of the lession was about 18 cm.

The only one abnormality in the laboratory tests was a 3- numeral ESR. The solid tumour was found in US examination and CT of the abdominal cavity forward to the aorta just above its bifurcation extending to the small pelvis. It was well limited, with heterogenous structure, without calcifications, with one decay focus. It extended probably from the anterior wall of the urinary bladder, adhered to the aorta's wall. The tumour's diameter was 18cm. The enlargement of lymph nodes was not found in the abdominal cavity. The cystoscopy and the contrast enema were normal.

The operation was done- the tumour extended from the ascendent part of the colon (diameter 14 cm). It was removed totaly with the appendix, the anterior part of the caecum, and with lymph nodes. The colon mucosa above the tumour was not changed.

There were no complication during the postoperative course. On the ground of pathologic examination we diagnosed: Angioleiomyosarcoma cum metastasibus ad lymphonodorum. In the abdominal cavity CT (done after the operation) enlargement of peritoneal lymph nodes remained as before.

The patient was treated with IX alternately used 5-days lasting courses of chemotherapy repeated every month, consisting of: ifosfamide (1800 mg/m^2) + vepeside (100 mg/m^2) and ifosfamide (1800 mg/m^2) + adriamycine (40 mg/m^2) and 10 times administration of vincristine (2,0 mg/m^2).

The boy achieved complete remission after the second cycle of chemotherapy. Now, he remains in the first complete remission 7 months after the end of therapy.

DISCUSSION

Angioleiomyosarcoma belongs to very rarely diagnosed malignant neoplasms, consisting of smooth muscular and vascular elements. The alone of leiomyosarcoma in solid tumours in adults is about 7%, but among muscular neoplasms angioleiomyosarcoma constitutes only a small percentage (Silvereberg, 1989). Only about 100 cases in adults have been

described till now (Schlittenbauer, 1996). It is mainly found in persons in middle and old age, exptionatly in hte childhood. Its most often localization in the peritoneum, superficial-subcutaneous regions ans blood vessel walls (Silverberg, 1989). Sometimes, it is found in the gastrointestinal tract, genitourinary or respiratory tracts, where it arises from smooth muscles found in the blood vessels (Vogel 1986; Pavlovich, 1989). The prognosis is univocally poor. The highest percentage of failures- 50-100% deaths during the short time of follow-up (Silverberg, 1989)- occurs when the tumour is localized in the peritoneal mesentery- like in our patient.

REFERENCES

Pavlovich A J, Bokarev M J - "A case of surgical treatment of angioleiomyosarcoma of the mediastinum" - Klin. Med. Mosk. (1989); 67 (10): 124-125.

Schlittenbauer M, Muller K M - "Angio-leiomyosarcoma of the lung- primary tumor or meta-stasis?" - Pneumologie, (1996); 50 (2): 215-228.

Silverberg S -"Soft tissue sarcomas: Histological diagnosis" - Ravenpress, (1989): 63-76.

Vogel R - "Rare tumors of childhood" from „Pediatric oncology and heamtology" - Hockenberry M J, Coody D K ed; the C V Mosby Comp., (1986): 160-165.