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Case Report

Spinal metastasis of nephroblastoma: Yes it exists [☆]

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ABSTRACT

Nephroblastoma or Wilms' tumor is the most common malignant tumor of the kidney in the pediatric population. Metastasis is caused by hematogenous spread. The most common localizations in decreasing order of frequency are lymph nodes, lungs, and liver. The bone is very rarely affected. According to the literature, bone metastases have been described in the iliac bone, skull, and mandible. The vertebral localization was described in 3 cases only, the first 1 in 2009, and the 2 others in 2015. The goal of our work is to report a very rare case of metastatic vertebral localization of a Wilms' tumor in relapse after treatment; and thus to underline the potential for vertebral and intracanal involvement in nephroblastoma.

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Case report

We report the case of a 10-year-old child, who was treated for left renal nephroblastoma in our hospital. The history of the disease goes back to 1 year, with the onset of abdominal pain. The initial evaluation revealed a prerupture stage 2 nephroblastoma without secondary localizations. The patient was initially put on preoperative chemotherapy for 4 weeks. The follow-up examination after 1 month was in favor of an increase in volume of the initial mass with the appearance of a right posterobasal pleural mass related to

secondary pleural localization. The decision after multidisciplinary staff was to resect the renal tumor by total enlarged right ureteronephrectomy without adrenalectomy, to proceed to second line chemotherapy type ICE (ifosfamide [Ifex], carboplatine and etoposide) 2 to 4 and a radiotherapy 25.2 Gy on the operating bed in 14 fractions of 1.8 Gy. Histology revealed a nephroblastoma with 3 components (Figs. 1 and 2): a blastematosus component (79%), anaplastic, and an epithelial component estimated at 21%. There was no mesenchymal component. The mass was high risk tumor. It was classified as stage III of the modified protocole of SIOP (international society of paediatric oncology). A mesenteric lymph node metas-

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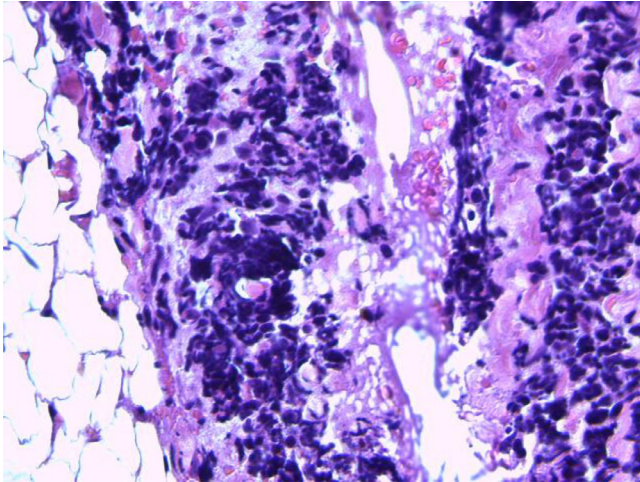


Fig. 1 – HE , Gx40 : infiltration of pleural and fatty tissue by the tumor.

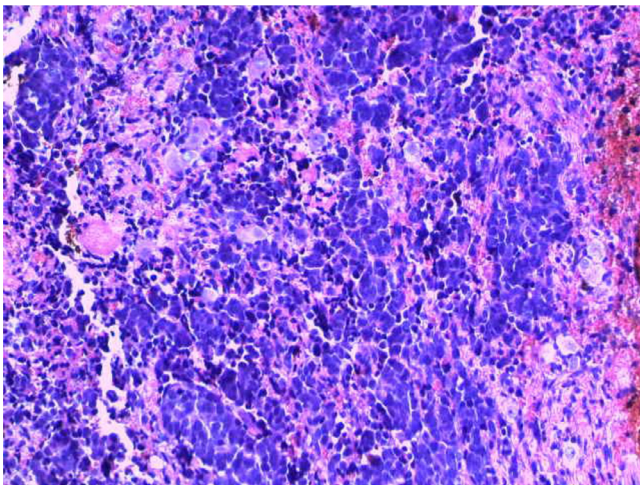


Fig. 2 – HE , Gx20 : blastomatous component of a metastatic nephroblastoma on pleuro-pulmonary.

tasis was associated. The control after the end of chemotherapy showed an increase in volume of the pleural mass. Therefore, a resection of the pleural mass was performed after W32 of the chemotherapy. The patient was referred for radiotherapy with second line chemotherapy type ICE (ifosfamide [Ifex], carboplatine and etoposide) 2 to 4. Three months later the patient developed severe dorsal-lumbar pain. The dorso-lumbar MRI showed a T1 hypo signal of the T11 and T12 vertebral bodies, enhanced after injection of gadolinium, a compression with recoil of the posterior wall of the T11 vertebral body in connection with secondary vertebral localizations (Fig. 3). A

right posterobasal pleural metastasis with endocanal extension and anterior epiduritis is associate (Fig. 4).

Discussion

Nephroblastoma or Wilms' tumor is a malignant renal tumor of young children. It is a tumor of the embryonic renal blastema. Renal tumors represent 6% of all cancers pediatrics [1] of which 90% are nephroblastoma. It occurs between 1 and 6 years with an average age of 3 years. It is rare after 7 years and exceptional after 15 years. According to the studies, More girls than boys develop Wilms' tumor. This type of tumor is usually unilateral but can be bilateral in about 5% of cases. Wilms' tumor is in 89% of cases sporadic, in 10% of cases a genetic predisposition and in the other 1% of cases familial.

The known metastases are the regional lymph nodes, liver, and lung. The bone extension represents 13% to 14% according to the literature [2]. Vertebral localization is very rare. According to the studies, there is no relation between the histological type of the initial tumor and the possibility of vertebral involvement [1]. In the Cohn study, vertebral metastases occurred in patients despite favorable histology [3]. They are the most frequent cause of intramedullary metastasis, which is associated with a very high mortality risk [2,4]. According to the studies, spinal cord compression is an exceptional event. In the Cohn et al. and Watanabe et al. studies, only 30 patients over a 42-year period had such an injury [3]. Similarly, in the Sudour-Bonnange H el ene study, 9 patients over 43 years were identified [1].

Bone locations of nephroblastoma can be asymptomatic [5] and should therefore be systematically searched for during extension evaluations. In the literature, bone metastases have been reported in the iliac bone 5, skull, and mandible [6]. On the radiological aspect and according to the publications reporting vertebral metastases, which are very rare, there are only 2 [1,2]. Lesions are often lytic, poorly limited 5 appears as a T1 hypo signal enhanced mass after injection with scintigraphic fixation [2]. In our case it is also a lesion of the vertebral bodies hypo T1 signal containing nodular lesions enhanced in periphery after injection (Fig. 2).

The therapeutic management of Wilms' tumor with vertebral metastasis and spinal cord compression is variable and intensive, including polychemotherapy, surgery of the renal mass and secondary localizations, and radiotherapy. The diagnosis must be made urgently in front of any evocative clinical symptomatology as it is the case of our patient. Decompression surgery is the rule in cases of spinal cord compression. Unlike other tumors where chemotherapy is more successful [1]. After the surgery, the specimen should be sent immediately for anatomopathological study with immunohistochemistry to eliminate other histological types such as juvenile renal cell carcinoma or rhabdoid tumor.



Fig. 3 – MRI images in sagittal section: (A) T1 sequence, (B) T2 sequence, (C) injected T1 sequence showing: T1 hyposignal of the vertebral bodies of D11 and D12 which are enhanced after injection of Gadolinium. The latter is the site of 2 nodular signal anomalies enhanced in the periphery. A biconcave compression fracture D11 with posterior wall recession and angulation of the spine.

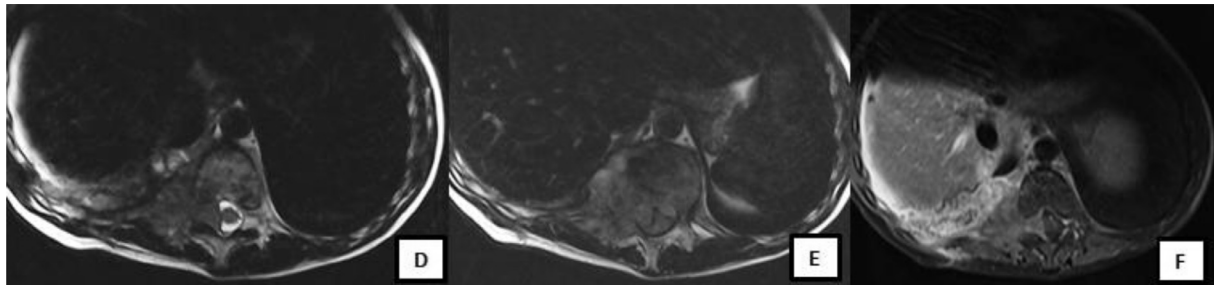


Fig. 4 – MRI images in axial section: (D) T2 sequence, (E) T2 sequence, (F) injected T1 sequence showing: Right posterobasal pleural mass with intermediate T2 signal, T1 hyposignal, enhanced after injection of Gadolinium. This mass: Invades the transverse processes D10-D11-D12 and their foramina with endocanal extension amputating the subarachnoid spaces and compressing the medulla which is displaced to the left without any signal abnormalities in the medulla. It extends to the right paravertebral muscles and the soft tissues in front of it.

Conclusions

In conclusion, vertebral and intramedullary metastases in Wilms' tumors are rare, but must be systematically sought during extension and control examinations.

Author's contributions

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

Patient consent

Written informed consent for publication was obtained from patient's parent.

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