Malignant peripheral nerve sheath tumor (MPNST) arising from an adrenal gland ganglioneuroma: A case report

F. Kediha, A. Zammouchi, M. Guermi, M. Amtout, N. Moulaï, W. Ouahioune

Department of Pathology, Frantz Fanon Teaching Hospital of Blida, Algeria

ABSTRACT

Ganglioneuroma is a benign tumor arising from the neural crest cells. This tumor is rare compared to other benign neural tumors such as schwannoma or neurofibroma. Most ganglioneuromas are diagnosed in young patients and are most often located in the posterior mediastinum followed by the retroperitoneum.

Ganglioneuromas are benign tumors rarely differentiate into malignant, and differentiation into malignant peripheral nerve sheath tumors (MPNST) is extremely rare. To date, there are less than 20 reported cases of MPNST arinsing from ganglioneuroma in the literature

We report a case of 24-year-old male with no history of childhood neuroblastoma or a familial neurofibromatosis, who presented with an adrenal gland mass discovered radiologically in the context of an exploration of low back pain.

Our finding demonstrated that, although rare, spontaneous malignant transformation of ganglioneuroma into MPNST can occur.

KYES WORDS: MPNST, ganglioneuroma, adrenal gland, young man

INTRODUCTION: Ganglioneuroma is a benign tumor arising from the neural crest cells; it contains no immature neuroblastic elements. This tumor is rare compared to other benign neural tumors such as schwannoma or neurofibroma. Most ganglioneuromas are diagnosed in patients between the ages of 10 and 40 years, and are most often located in the posterior mediastinum followed by the retroperitoneum.

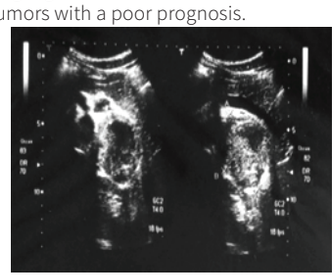
Ganglioneuromas are benign tumors rarely differentiate into malignant, and differentiation into malignant peripheral nerve sheath tumors (MPNST) is extremely rare and scarcely reported. To date, there are less than 20 reported cases of MPNST arising from ganglioneuroma in the literature.

Most MPNSTs are highly aggressive tumors with a poor prognosis.

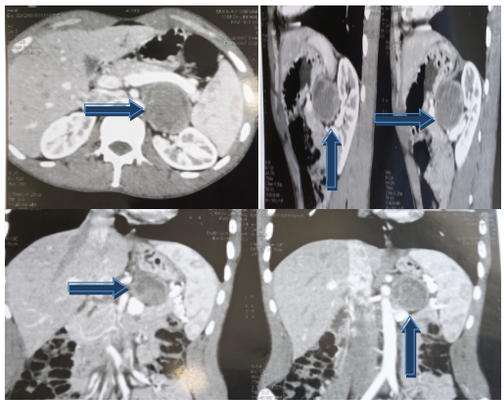
OBSERVATION: We report the case of a 24-year-old man with no history of childhood neuroblastoma or previous radiation therapy or a familial neurofibromatosis, who presented a lower back pain for about 04 months.

The radiological examination shows a well-limited retroperitoneal mass, measuring (62x48) mm, located near the left renal pedicle.

Surgical resection was done.



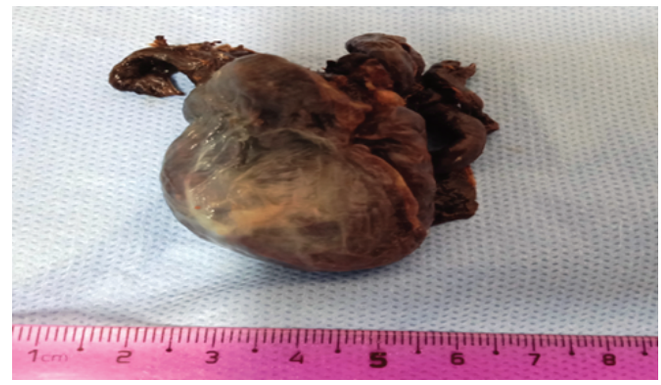
Imagery : Voluminous hypoechogenic mass



Imagery : Well-limited retroperitoneal mass, measuring (62x48)mm, located near the left renal pedicle

RESULTS

MACROSCOPY: We received an encapsulated nodular mass measuring (7x5x3,5) cm, partially surrounded by fat and an adrenal gland measuring (2,5x2,5) cm Cut section shows a multilocular cystic appearance. Cysts were filled with hemorrhage and necrosis.

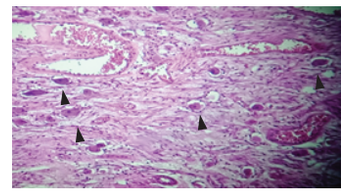


Macroscopy: Encapsulated nodular mass partially surrounded by fat (arrow)

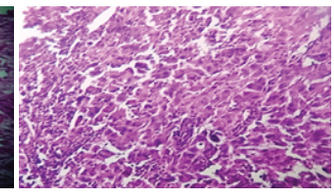
HISTOPATHOLOGY: Histological examination shows two well demarcated tumor components: benign and malignant.

The benign component corresponds to a ganglioneuroma, consisted of mature ganglion cells scattered throughout a schwannian background.

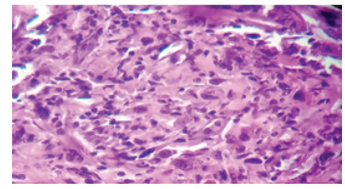
The malignant component is composed of clusters of strongly atypical cells of medium to large size. The cells are spindle-shaped focally and multinucleated giant cells are present. The mitoses are numerous. Larges foci of necrosis and hemorrhage are observed.



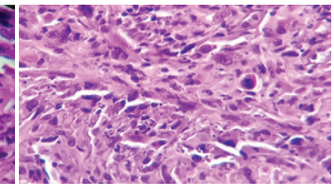
HE: Ganglioneuroma (ganglion cells "arrow" in a schwannian background)



HE: Sarcomatous component



HE: Epithelioid cells in the sarcomatous component



HE : Atypical mitosis (arrow)

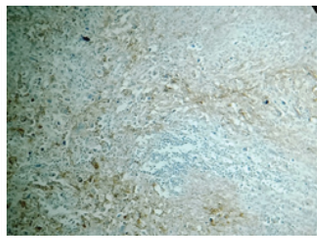
IMMUNOHISTOCHEMISTRY:

Cells of the malignant component (sarcomatous) focally express PS100, NSE, CD56 and EMA

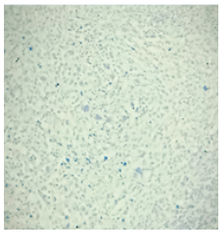
The other markers used (Chromogranin A, Synaptophysine, Melan A, AE1/AE3, CD20, CD3, CD163, Desmine, CD34) are all negative

The diagnosis retained is a MPNST occurring on a ganglioneuroma of the adrenal gland.

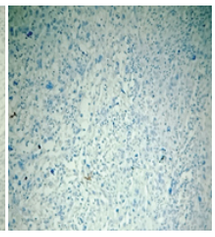
IHC: positive staining for PS100



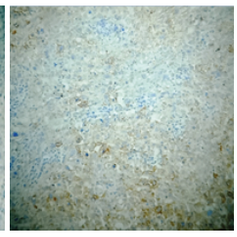
IHC : Positive staining for EMA



IHC: Negative staining for synaptophysine



IHC: Negative staining for chromogranine A



IHC: Positivre staining for CD56

COMMENTS: Ganglioneuromas are the rarest and most benign of the neuroblastic tumors [1].
They are composed of mature ganglion cells and Schwann cells [1].
The most affected sites are the posterior mediastinum, retroperitoneum, adrenal gland and soft tissue of the head and neck [2]. In our case the lesion was located in the adrenal gland.
The majority of ganglioneuroma manifest as an asymptomatic mass, which are discovered on routine radiographic studies for other lesions [4]; the present case was symptomatically and presented with a lower back pain.
Despite its extremely rare incidence, malignant transformations may occur and the neoplasm may exist in the form of MPNST [4].
Some studies reported that a few patients had a history of radiotherapy or neurofibromatosis but many studies report that the occurrence of malignant schwannoma in the absence of familial neurofibromatosis is extremely rare [5].
The present case has no evidence of familial neurofibromatosis or radiation history, indicating that other factors may involved.
Studies on how MPNST may be derived from ganglioneuroma are required to elucidate the mechanisms of this transformation; the most likely initiator is the Schwann cell [6].
Surgical resection is the primary treatment of MPNST, which is usually followed by a poor outcome [7]. Our patient received a complete surgical excision.
The diagnosis of MPNST is difficult, it depends on the pathology and immunohistochemistry examinations [5].
The prognosis of MPNST has been poorly reported [8]. MPNST is resistant to radiation and chemotherapy, so there are no effective therapeutic methods to be used postoperatively.

CONCLUSION

The transformation of a ganglioneuroma into a malignant peripheral nerve sheath tumors (MPNST) is extremely rare.
These tumors generally occur in young patients.
The mechanism of transformation is unknown and the prognosis is poor.
In the perspective of our case, although extremely rare, further investigations and studies must be made to elucidate the mechanisms of the transformation of ganglioneuroma into a MPNST.

REFERENCES

1- Enzinger and Weiss's SOFT TISSUE TUMORS SIXTH EDITION; p792-795
2- Christopher D.M Fletcher; Diagnostic histopathology of tumors, Forth Edition/Volume 1, MD, FRCPath, p1290
3-Christopher D.M Fletcher, Julia A. Bridge, Pancras C.W Hogendoorm, Fredrik Mertens, editors. WHO Classification of Tumors of Soft Tissue and Bone; 4th Edition, 2013, p187
4- Nicole Berger-Angela Borda ; Pathologie thyroïdienne, parathyroïdienne et surrénalienne; p350-351
5- Z.H. MENG, Y.S. YANG, K.L. CHENG, G.Q. CHEN, L.P. WANG, and W. LI; A huge malignant peripheral nerve sheath tumor with hepatic metastasis arising from retroperitoneal ganglioneuroma ; 2012;
6-Jean-Pierre De Chadarévian, Judy MaePascasio, Gregory E. Halligan ; Malignant Peripheral Nerve Sheath Tumor Arising from an Adrenal
Ganglioneuroma in a 6-Year-old Boy; First Published May 1, 2004 Research Article Find in PubMed: https://doi.org/10.1007/s10024-004-8084-9
7-G Drago , B Pasquier, D Pasquier, N Pinel, V Rouault-Plantaz, J F Dyon, C Durand, C Armari-Alla, D Plantaz; Malignant peripheral nerve sheath tumor arising in a "de novo" ganglioneuroma: a case report and review of the literature DOI: 10.1002/(si-ci)1096-911x(199703)28:3<216::aid-mpo13>3.0.co;2-c
8-S Damiani , V Manetto, G Carrillo, A Di Blasi, O Nappi, V Eusebi; Malignant peripheral nerve sheath tumor arising in a "de novo" ganglioneuroma. A case report; Institute of Anatomic Pathology, University of Bologna, Italy. PMID: 1850181