

# Cystic Nodular Ganglioneuroblastoma: Case Report

Ganglioneuroblastoma nodular quístico: Presentación de un caso



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## Key words (MeSH)

Ganglioneuroblastoma  
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## Palabras clave (DeCS)

Ganglioneuroblastoma  
 Glándulas suprarrenales  
 Niño

## Summary

We present the case of a child patient aged 9 years, in whom, as an incidental finding in chest X-rays, a calcified lesion was observed in the left hypochondrium. The study is extended by ultrasound and then MRI was performed to better characterize it. These showed a left adrenal, predominantly cystic mass with a solid nodule. The surgical team decided to perform resection of the mass and submit it for histopathological examination. A definitive diagnosis of cystic nodular ganglioneuroblastoma was made. Ganglioneuroblastoma is a rare tumour, originating in ganglion cells of the sympathetic nervous system. The site of more frequent origin is in the adrenal glands. Although the majority are diagnosed based on the postoperative histological analysis, with ultrasound and MRI we should include it among the differentials diagnoses of adrenal incidentalomas in a child or young patient. The findings on CT are variables.

## Resumen

Presentamos el caso de un niño de 9 años de edad, en quien como hallazgo incidental en una radiografía de tórax se observa una calcificación en el hipocondrio izquierdo sugestiva de masa. Se amplía el estudio mediante ecografía y resonancia magnética (RM) abdominal para mejor caracterización: se visualiza una masa suprarrenal izquierda, predominantemente quística, con un nódulo sólido. Finalmente, se le practica una resección quirúrgica, en la cual se llega al diagnóstico de ganglioneuroblastoma nodular quístico, tras el análisis histopatológico. El ganglioneuroblastoma quístico es un tumor poco frecuente, originado en las células ganglionares del sistema nervioso simpático. El sitio de origen más frecuente son las glándulas suprarrenales. A pesar de que el diagnóstico definitivo se suele realizar tras la resección quirúrgica del tumor, con la ecografía y la RM se debe incluir entre los diagnósticos diferenciales de incidentalomas suprarrenales en un niño o un paciente joven. Los hallazgos en tomografía computarizada son variables.

## Introduction

Ganglioneuroblastoma is a rare tumor that originates in the ganglion cells of the sympathetic nervous system. Most of them have been observed in children under 5 years of age. The most frequently affected organ is the adrenal glands.

The vast majority of published cases describe it as an adrenal incidentaloma in studies carried out for other reasons, since the associated clinic is nonspecific: they may be associated with abdominal pain or distension when they are large.

Imaging techniques, especially ultrasound and magnetic resonance imaging (MRI), are of great help in the characterization of injuries, especially

in a child or young patient, as they do not radiate and are relatively easy to access. Although these techniques can be used to approximate the definitive diagnosis, in the vast majority of cases the definitive diagnosis is made by histopathological analysis of the anatomical piece obtained in the surgical resection.

## Clinical case

Male patient aged 9 years who consults the emergency department for a respiratory infection clinic, consisting of cough, expectoration and fever



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at 39 °C. Does not refer to aboveclinical teeth. A chest x-ray is initially performed, and a blood count and CRP are taken. Blood count shows a slight leukocytosis of 13000 with neutrophils of 85% and PCR is discreetly elevated, 20 units. In the chest x-ray, no consolidation of the airspace is observed, only a small thickening of the peribronovascular interstitium. However, as an incidental finding, there is a calcification in the left hypochondrium, of curvilinear morphology (Figure 1), which suggests that it is a calcified mass, so it is recommended to complete study with other imaging techniques. The patient is discharged with analgesic, antipyretic and antihistamine treatment. An abdominal ultrasound scan is performed, showing a left adrenal lesion with mostly cystic contents and a solid inner pole (Figure 2). To better characterize the fin-

dings, an MRI is performed, where it is observed that the adrenal lesion is mostly cystic, with hemorrhagic areas that produce liquid-liquid level (Figure 3). The wall of the mass is also calcified, as shown on the chest x-ray. After administration of intravenous gadolinium, nodular enhancement is observed on the more caudal side of the lesion (Figure 4), which in the sequence shows restriction of diffusion (Figure 5). In endocrine blood tests, catecholamines and hormone levels are normal. A differential diagnosis of the mass between a hematoma or a cystic tumour of the neuroblastoma or less likely teratoma type is considered.

The pediatric surgery team performs the resection of the mass. Histopathological analysis diagnoses nodular cystic ganglioneuroblastoma, rich in stroma.

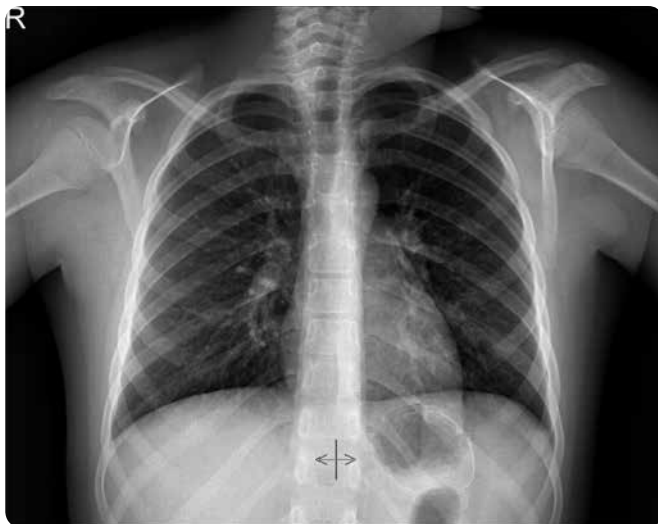


Figure 1. Chest X-ray. Discrete thickening of the peribronovascular interstitium bilaterally. As an incidental finding a calcification of curvilinear morphology in the left hypochondrium, suggestive of mass.



Figure 2. Abdominal, sagittal ultrasound of the left hypochondrium. A mass in the left adrenal gland of heterogeneous ecogenicity, predominantly hypoechoic with what appears to be a solid pole within.

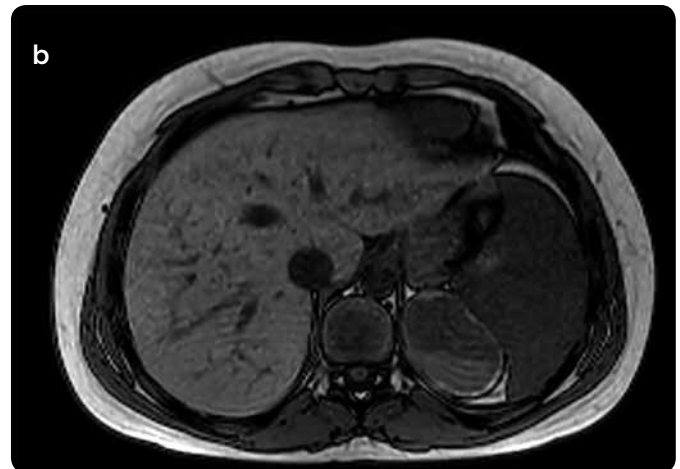
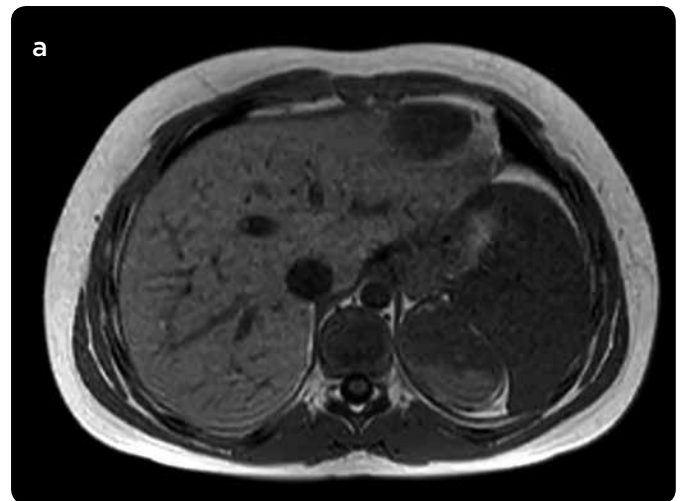


Figure 3. a) Axial image of MR enhanced in T1 in phase and b) out of phase. The left adrenal mass is mostly cystic, with a high signal flange corresponding to the peripheral calcification, visualized on the chest x-ray. Liquid-liquid level, with the lowest level of high signal hemorrhage. There is no macroscopic fat or signal drop in the out-of-phase sequence suggesting microscopic fat in the lesion. It has no greasy component.

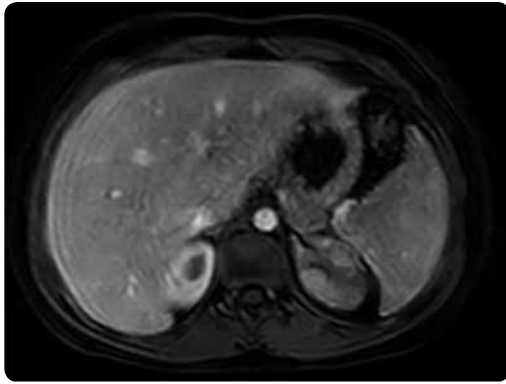


Figure 4. Axial image of MRI, with contrast dye in portal phase. The mass shows solid poles that are enhanced with the administration of intravenous contrast dye.

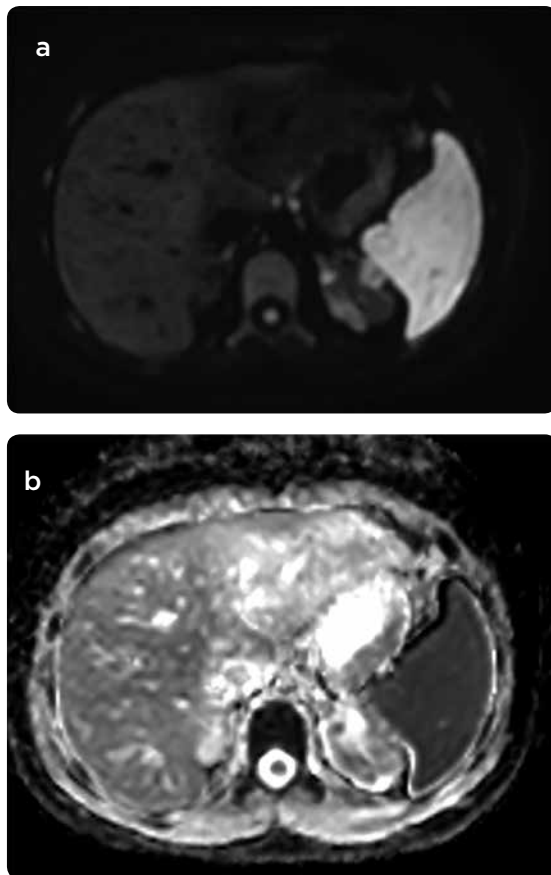


Figure 5. a) Axial MR diffusion image ( $b=1000 \text{ sec/mm}^2$ ) and b) on ADC map. Diffusion restriction is shown at the solid poles, with low ADC values of  $0.95 \times 10^{-3} \text{ mm}^2/\text{sec}$ .

## Discussion

Ganglioneuroblastoma is a rare tumor that originates in the ganglion cells of the sympathetic nervous system. More than 90% have been observed in children under 5 years of age and are considered a variant of neuroblastoma. In histopathological analysis they may show a nodular or diffuse pattern, but the nodular pattern is more aggressive, and can be metastasized.

The most frequent site of origin is the adrenal glands, as in this case; it has also been observed in the retroperitoneum, in the posterior mediastinum and, less frequently, in the neck (1). The malignant potential of ganglioneuroblastoma is intermediate between neuroblastoma and ganglioneuroma (2). Clinical findings are usually nonspecific, sometimes causing abdominal pain or distention from the mass effect, when they are large.

It may be seen as an adrenal incidentaloma and is difficult to tell the difference from other adrenal masses. For example, on CT it may have several appearances, such as solid or predominantly cystic, or with some solid pole, relative to the number of ganglionic tumor cells. MRI is superior for diagnosis and, above all, useful in the detection of metastasis. Ganglioneuroblastoma is a low signal mass in sequences with T1 information and high signal mass in sequences with T2 information. In the dynamic study, with intravenous contrast dye administration, it shows early enhancement, in contrast to ganglioneuroma (3). However, it is difficult to reach a definitive pre-surgical diagnosis with the image, so most of the time it is post-surgical.

Nuclear medicine studies are usually not indicated, except in endocrine active lesions (4), in catecholamine-producing tumors and to rule out metastasis.

It should be noted that in this case the initial finding was visualized on the chest x-ray, hence the importance of using an appropriate reading system, otherwise it would not have been detected. However, if a predominantly cystic adrenal incidentaloma is found in a child or young patient, neuroblastoma, among others, such as neuroblastoma, ganglioneuroma, pheochromocytoma, teratoma and less frequently carcinoma, should be included in the list of differential diagnoses. Cystic neuroblastomas are non-functional tumours, usually with calcifications and added hemorrhagic component (5). Ganglioneuroma is a rare benign tumor detected as incidentaloma. They are usually large, larger than 20 cm, and the findings in the image are nonspecific (6). Adrenal teratomas are rare, but it is a typical location in children, and solid, cystic, or mixed-component imaging studies may appear (7).

It should be noted that cystic lesions of the adrenal glands can be grouped into three categories: pure cystic, parasitic and solid pole cysts, the latter associated with bleeding or necrosis.

In general, purely cystic lesions correspond to simple cysts, pseudocysts, lymphangiomas or hemangiomas. Adrenal cysts are rare with an incidence of 5.7 % of incidentalomas (8).

Cysts with hemorrhage are associated with perinatal stress sequelae, traumatic etiology is less accepted. Hydatid cysts in the adrenal glands are very rare, not a typical location (9).

In addition to imaging studies, it is always necessary to establish whether the lesion is hormone-secreting or not in the event of an adrenal incidentaloma, since the management is different for each entity (10).

Treatment is surgical and sometimes requires chemo and radiation therapy, although there is no general consensus. There are insufficient follow-up data over long periods of time

## Conclusion

Cystic nodular ganglioneuroblastoma is a rare tumor of the ganglion cells of the sympathetic nervous system, which most often originates in the adrenal glands.

Although the definitive diagnosis is obtained after histopathological analysis, imaging techniques such as ultrasound and MRI allow the characterization of the components of the lesion to make an approximation to the diagnosis. The findings on CT are variable and there are no conclusive data to establish their contribution.

Systematic chest x-ray reading is a very useful tool and should be done routinely, as this case demonstrates.

In the case of an adrenal incidentaloma, the age of the patient and the main component of the lesion should be observed, which reduces the diagnostic possibilities.

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