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The Mediastinal Ganglioneuroma in a Case Operated on in Mali

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ABSTRACT

Ganglioneuroma is a benign tumor of the sympathetic nervous system belonging to the neuro-cristopathies, the pathogenesis of which remains less well known. In Mali, no mediastinal ganglioneuroma has been published in the literature. The diagnosis was confirmed by anatomopathological examination. We report a case of mediastinal ganglioneuroma in a 14-year-old female patient and review the literature.

Keywords

Ganglioneuroma, Mediastinal, Surgery, Mali.

Introduction

Ganglioneuroma is a rare, well-differentiated, benign neurogenic tumor that develops from mature sympathetic ganglion cells and nerve fibers of thoracic origin. It arise from neural crests (like neuroblastomas and ganglioneuroblastomas). It is a rare tumor, constituting 0.07 to 0.2% of tumors. Their starting point is the sympathetic chains that extend from the base of the skull through the neck, posterior mediastinum and retroperitoneum to the adrenal glands. Children and adolescents are preferentially affected with a slight female predominance. The prognosis of this tumor is related to early diagnosis and complete or incomplete excision, the pathologist is the cornerstone of the diagnosis and histological type.

Observation

Patient M. S, 14 years old with no known medical-surgical history, admitted for exertional dyspnea stage II according to NYHA associated with intermittent cough and right basi-thoracic pain radiating to the back calmed by usual analgesics requiring a medical consultation at the Center for Cardiovascular Surgery ANDRE FESTOC of Bamako. On admission: he was in good general condition with colored conjunctiva. He had no cyanosis or digital hippocrasis. Pleuropulmonary examination revealed a right basal pulmonary condensation syndrome. Heart sounds were normal. The rest of the examination was unremarkable. The chest X-ray showed a para-cardiac opacity (Figure 1). The thoracic CT scan (Figure 2) showed a large mediastinal mass measuring latero-posterior and enveloping the large vessels (right pulmonary artery and aorta). The biological workup was normal. In view of this picture, an exploratory thoracotomy was decided and performed. She was admitted to the operating room under general anesthesia with selective intubation through the Carlens tube. A posterior lateral thoracotomy passing the 5th right intercostal space was performed. The surgical exploration showed a large mass of firm consistency, grayish color, developing in the posterior mediastinum of the 4th, 5th, 6th and 7th intercostal nerves and then the sympathetic nerve (Figure 3) and poorly vascularized. We decided to perform a biopsy by taking three cores. Surgical excision was not performed because of the envelopment of the surrounding structures, putting the vital prognosis at stake. We inserted two CH 32 thoracic drains including an apical and posterior drain. The

postoperative course was simple. The histology result was in favor of a benign mediastinal ganglioneuroma.

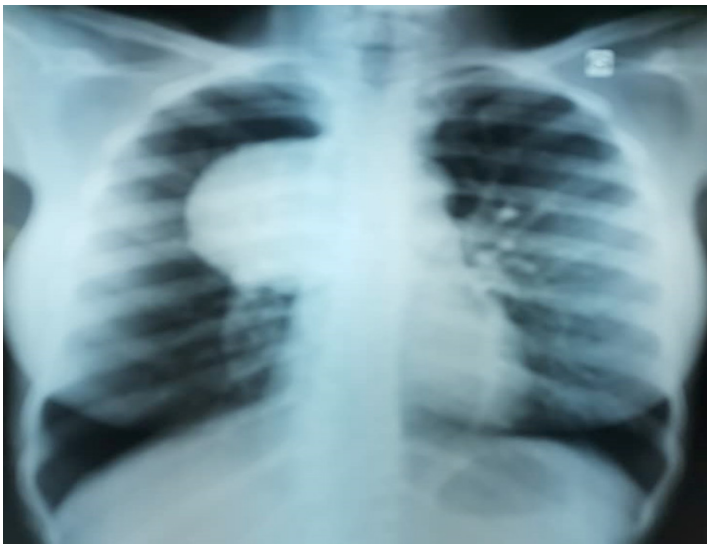

Figure 1: Chest X-ray showed a large para-cardiac opacity with the silhouette.

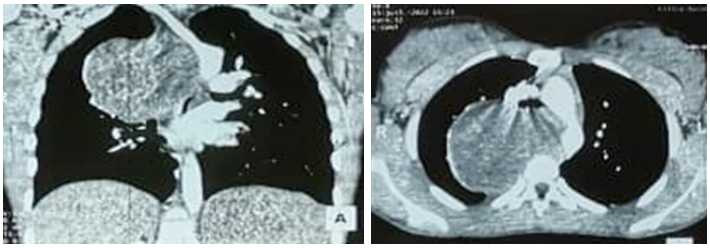

Figure 2: Chest CT scan showing a large tissue-like mass enveloping the grox vessels, trachea and esophagus.

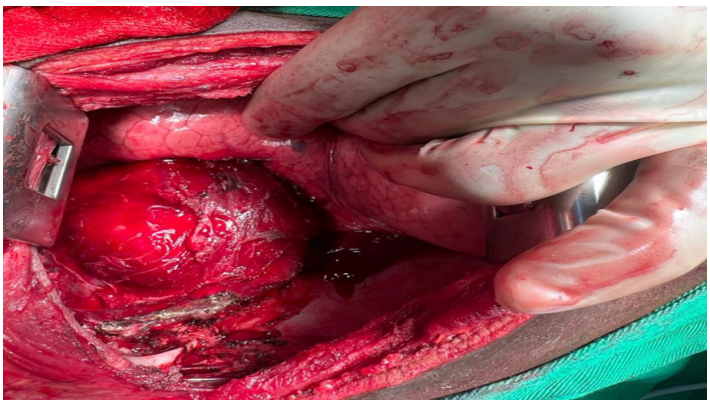

Figure 3: Preoperative view of the mass.

Discussion

Ganglioneuroma is a rare benign tumor of neuroectodermal origin. This tumor develops, like neuroblastoma and ganglioneuroblastoma, from the sympathetic nervous system [1,2]. It is composed of mature ganglion cells and a stroma containing nerve cells and a schwanian contingent, whereas neuroblastoma and ganglioneuroblastoma are immature lymph node cells that have a greater potential for progression [3]. The ganglioneuroma develops along the sympathetic chain from the neck to the pelvis. Our patient has a mediastinal ganglioneuroma which corresponds to the development of the sympathetic chain. This mediastinal localization is the most frequent after the retroperitoneum [4,5]. It occurs at all ages, but is mostly found in children and young adults [6].

Females are more often affected with a sex ratio of 0.75. These tumors evolve quietly and are most often discovered by chance during a radiological examination for another disease. The radiological diagnosis of these tumors is difficult. However, imaging allows the location of the tumor to be determined as well as its relationship with the surrounding organs, particularly the vessels. Ultrasound is not very specific and often reveals a heterogeneous, well-defined tissue mass in the adrenal cavity. The tumor may come close to the vessels without invading them [7].

The CT scan is aspecific. Anatomically, the ganglioneuroma reproduces the architecture of a sympathetic ganglion [8]. Macroscopically, it is an oval, encapsulated, fibrous, whitish-gray, firm tumor with occasional calcifications. The radiological diagnosis of these tumors is difficult. However, imaging allows the location of the tumor to be determined as well as its relationship with the surrounding organs, particularly the vessels. Ultrasound is not very specific and often reveals a heterogeneous, well-defined tissue mass in the adrenal cavity. The tumor may come close to the vessels without invading them [7,9].

The CT scan is aspecific. In our patient, the CT scan showed a large mass in the upper mediastinum surrounding the large vessels which was not specific. The hormonal secretion test was not performed in our patient but the ganglioneuroma is usually a non secreting tumor. Nevertheless, the diagnosis of certainty will only be made after a histological study of the surgical specimen. Indeed, although preoperative biopsy allows the diagnosis to be made, a complete analysis of the excisional specimen is still necessary because of the possibility of contingents of neuroblastoma but also of pheochromocytoma within the ganglioneuroma [8,9].

The treatment remains surgical. It consists of tumor removal; this procedure is all the more difficult as the tumor is large and has intimate relationships with the neighboring structures, in particular the large vessels (IVC and aorta). This total removal, which is the curative treatment, was not performed because of the surrounding structures, but this thoracotomy made it possible to perform a biopsy on the patient.

Conclusion

The ganglioneuroma is a rare benign tumor developed from a mother cell of the neural crest. The most frequent site is mediastinal followed by retroperitoneal. The prognosis of this tumor is related to early diagnosis and complete excision. The diagnosis is purely histological as in our patient.

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