American Academic Scientific Research Journal for Engineering, Technology, and Sciences
ISSN (Print) 2313-4410, ISSN (Online) 2313-4402
http://asrjetsjournal.org/

Anemic Syndrome Revealing Rhabdomyosarcoma of the Right Heart: About Clinical Case with Literature Review

S. Bellouize[a*], N. Loudiyi[b], A.Moujahid[c], Y.Bekkali[d], A.Benyass[e]

[a,d]*Cardiac surgery department of Mohammed V military hospital of Rabat Morocco*
[b,e]*Cardiac department de cardiology of Mohammed V military hospital of Rabat Morocco*
[c]*Cardiac intensive care department of Mohammed V military hospital of Rabat Morocco*
[a]*Email: lallasiham3@gmail.com*

Abstract

Rhabdomyosarcoma is one of the most common primary cardiac tumors in children and adolescents alongside lymphomas. The diagnosis is based on a bundle of clinical, biological and histological arguments. Cardiac imaging, dominated by MRI, plays a key role in the tissue characterization of masses Therapeutic management is based on chemotherapy and the most complete surgical resection possible. We report the case of a rhabdomyosarcoma of the right atrium with pulmonary metastases in a 16-year-old girl revealed by a severe anemic syndrome with hemoptysis.

Keys words: primitives' cardiac tumors; rhabdomyosarcoma; MRI; pulmonary metastases.

1. Introduction

Primary cardiac tumors in children and adolescents are rare, mostly benign. Only 10% are malignant, dominated by sarcomas (angiosarcomas, synovial sarcomas, rhabdomyosarcomas), but also primary lymphomas of the heart. Optimal management is essentially based on a precise anatomopathological diagnosis, associated with chemotherapy with the most complete surgical excision possible, on which the prognosis depends. We report the case of a rhabdomyosarcoma of the right atrium with pulmonary metastases in a 16-year-old girl revealed by a severe anemic syndrome with hemoptysis.

2. Observation

This is a 16-year-old patient, with no particular history, who has had a deterioration in her general condition for a month with exertional dyspnea and hemoptysis.

---

* Corresponding author.

Objective clinical examination of the discolored conjunctivae with clearly perceived heart sounds without murmur, but crackles at the bases of the lungs Chest x-ray found cardiomegaly with multiple bilateral alveolointerstitial opacities Biologically, there is an iron deficiency anemia at 6 g / dl. The CT scan of the chest revealed scattered nodular hyperdense lung lesions with clear borders evoking metastases with alveolar hemorrhage, associated with the presence of a parietal process of the right atrium measuring 35 mm in thickness extended over 65 mm reaching contact. from the mouth of the superior vena cava. Transthoracic echocardiography has objectified a dilated right auricle seat of a mass of heterogeneous structure and regular contour, measuring 24/34 mm, with invasion of the free wall of the RV, whose function is altered, associated with a pericardial effusion of low abundance, with good systolic function of the left ventricle at 60%. Cardiac MRI revealed the presence of a mass from the right atrium extended to the right ventricle, with regular outline, isointense on T1-weighted anatomical sequences, hyperintense on T2 STIR, with heterogeneous enhancement after injection of gadolinium. After blood transfusion and respiratory stabilization (Corticotherapy, antibiotic therapy and NIV sessions), the patient was operated. Per operatively, the heart is the site of a tumor of the right atrium extending towards he right atrioventricular sulcus and towards the superior vena cava measuring 7/5 cm. After a right atriotomy near the right atrioventricular sulcus, total resection of the tumor was performed and installation of a heterologous pericardial patch restoring the lateral wall of the right atrium. The histological and immunohistochemical study came back in favor of a rhabdomyosarcoma. The postoperative course was marked by alveolar hemorrhage requiring massive transfusions with vasoactive drugs and vasopressors. The evolution was marked by severe respiratory failure leading to death despite resuscitation maneuvers.

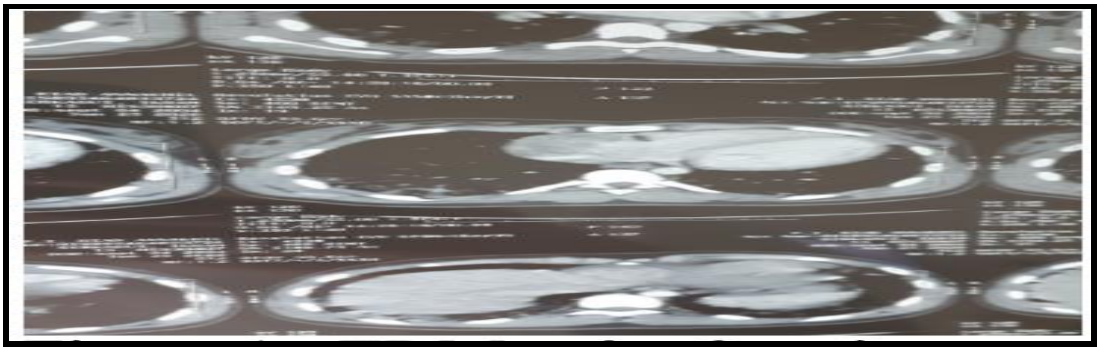

Figure 1: Preoperative CT scan, 4-cavity section. Tumor of the right auricle, developed at the expense of its free edge.

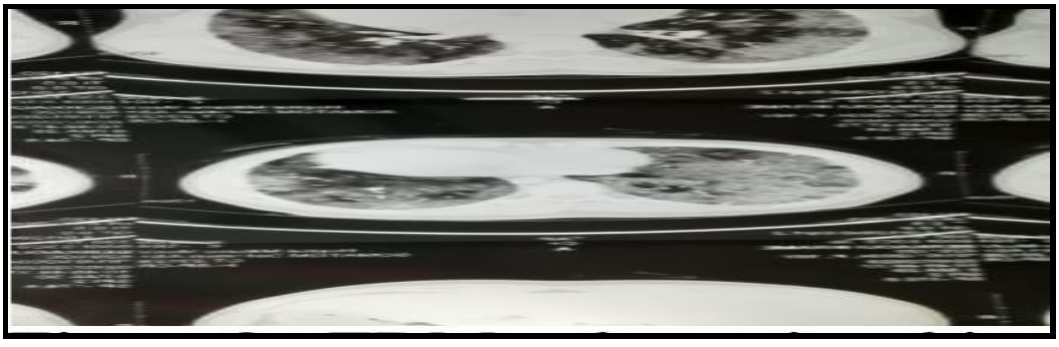

Figure 2: Pulmonary CT showing pulmonary metastases and alveolar hemorrhage.

Figure 3: Cardiac MRI: T1-weighted dark blood anatomical sequences.

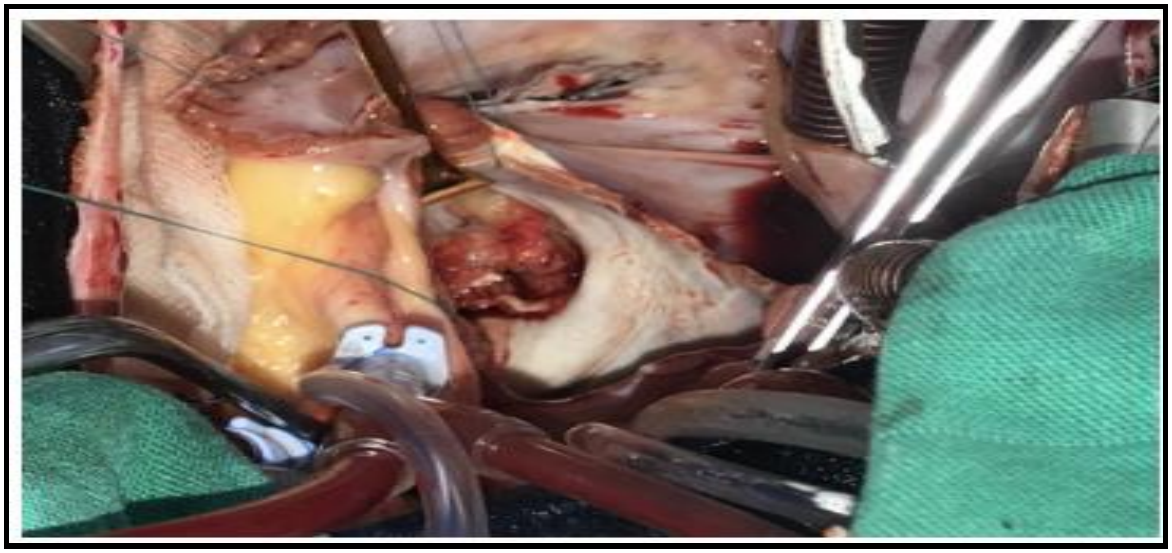

Figure 4: macroscopic appearance of the tumor of the right atrium during surgery.

3. Discussion

Cardiac tumors in children and adolescents represent a heterogeneous group of rare diseases, with an estimated prevalence between 0.0017 and 0.28%. [1,2]. These are most often benign tumors. Malignant tumors are either sarcomas or lymphomas. Primary cardiac tumors in children present a variety of clinical symptoms and are often revealed by locoregional complications (signs of heart failure, rhythm and conduction disorders, tamponade, pulmonary or systemic thromboembolic manifestations) or general (altered general condition, fever, arthralgia).

Trans thoracic echocardiography and trans esophageal echocardiography play an essential role in the diagnosis, making it possible to specify the location of the tumor, its relationships, and its characteristics (echogenicity´, heterogeneity´, dimensions, limits, infiltrating character). Cross-sectional imaging (CT scan and cardiac MRI) is the cornerstone both for diagnosis and for pre-therapeutic management thanks to the tissue characterization of the masses using the different image acquisition modes (anatomical blood sequences T1-, T2-weighted, fat-

suppression, and gadolinium injection) [3,4]. The diagnosis of malignant cardiac tumor of the heart is evoked on the clinic (symptomatic tumor in an older child or adolescent)

and imaging (heterogeneous, infiltrating, auricular or ventricular tumor). The vast majority of cases are sarcomas (angiosarcoma, synovialosarcoma, rhabdomyosarcoma.) [5], characterized by their infiltrating capacity, their high risk of metastatic dissemination and their particularly reserved prognosis. In a second step, depending on this clinic radiological analysis, biopsy by endovascular route or by thoracotomy can be discussed. It confirms the diagnosis of malignancy and specifies its histological type [6]. Therapeutic management depends on the histological type, mainly based on chemotherapy and surgery, which often remains difficult [6]. The prognosis is often guarded with a 1-year survival of 15% in adults, and few reported cases of surviving children [7].

4. Conclusion

Heart tumors in children and adolescents constitute a group of rare pathologies, heterogeneous in their histology, their therapy and their prognosis, but for which a precise diagnostic investigation is necessary, in order to propose the most adapted management.

References

[1] McAllister Jr HA. Primary tumors of the heart and pericardium. PatholAnnu 1979;14(Pt 2):325–55.

[2] Nadas AS, Ellison RC. Cardiac tumors in infancy. Am J Cardiol.

[3] Sparrow PJ, Kurian JB, Jones TR, et al. MR imaging of cardiac tumors. Radiographics2005;25:1255–76.

[4] Araoz PA, Eklund HE, Welch TJ, et al. CT and MR imaging of primary cardiac malignancies. Radiographics 1999;19:1421–34.

[5] Roberts WC. Primary and secondary neoplasms of the heart. Am J Cardiol 1997;1:671–82.

[6] primary cardiac tumors in childhood and adolescence Author links open overlay panel B.FresneauO.OberlinL.BrugièresD.Valteau-CouanetC.Patte 11 February 2010.

[7] Nakamichi T, Fukuda T, Suzuki T, et al. Primary cardiac angiosarcoma: 53 months' survival after multidisciplinary therapy. Ann ThoracSurg1997;63:1160.