

Giant Cervical Solitary Gardner Fibroma A Case Report and Review of the Literature

Zahra Sayad, Olaya Hamidi, Hafsa Elouazzani, Salma Benazzou, Nadia Cherradi and Malik Boulaadas
Faculty of Medicine and Pharmacy, Mohammed V University Rabat, Morocco.

ARTICLE INFO

Article history:

Received: 18 May 2021;

Received in revised form:
19 June 2021;

Accepted: 29 June 2021;

Keywords

Gardner Fibroma,
FAP,
Diagnosis,
Prognosis.

ABSTRACT

Gardner's fibroma is a rare benign soft tissue tumor most commonly occurring between infancy and adolescence. Approximately 70% are associated with familial adenomatous polyposis (FAP) and Gardner's syndrome which are inherited in an autosomal dominant manner. We report the observation of a young 16-year-old patient who underwent surgical resection of a large right lateral cervical mass, whose anatomopathological analysis of the operative specimen confirmed the diagnosis of Gardner's fibroma. Considering the association between this histological type and the FAP, a family survey and an endoscopic digestive exploration in search of a recto-colic polyposis coming back negative. The diagnosis is an isolated sporadic Gardner's fibroma.

© 2021 Elixir All rights reserved.

Introduction

Gardner's fibroma is a rare benign tumor in its sporadic form, as it is often seen with familial adenomatous polyposis (FAP) as part of Gardner syndrome. Its location includes the trunk, including the paraspinal region, head and neck.

It occurs mostly in children under 10 years old, less commonly in adolescents and young adults without gender predominance. (1,2)

However, screening for familial adenomatous polyposis in the diagnosis of Gardner's fibroid is suggested due to the association. The diagnosis is radiologically obvious but requires anatomopathological confirmation which remains to this day the only definite diagnosis. The imaging of choice is MRI. Surgery was the basis of treatment. Close monitoring by clinical and radiological examination is essential. (2,3)

The treatment and prognosis of Gardner's fibroid depend on the presence of PAF and other genetic conditions.

The goal of our work is to focus on this tumor in its isolated sporadic presentation especially at cervical location, to underline their clinical and histological aspects and the importance of a diagnostic and therapeutic follow-up.

Case a report

A young 16-year-old patient, with no medical family history or comorbidities, presented an enormous right lateral cervical swelling which had been evolving for 2 years, without associated signs. The clinical examination objectified a voluminous mass right lateral cervical polylobed, firm, painless, well limited, fixed in the deep plane, with a shiny aspect of the skin opposite and telangiectasias, measuring 15 cm long axis. The remainder of the cervicofacial examination was normal. The nasofibroscope was normal.

A cervicothoracic computed tomography scan with injection of contrast product showed multiple large, heterogeneous nodular formations, which come into contact with the brachiocephalic vein below, extend up to the height

of the vertebral body of C2, pushing back the right jugulocarotid axis and the tracheodigestive axis, with a reduction of the tracheal lumen in places. (Fig. 1)



Figure 1. Computed tomography in axial and coronal plans showing a giant lobulated lateral cervical mass, which is heterogeneously enhanced, exerting a mass effect on the vascular axis and the aerodigestive tract.



Figure 2. photo of the operative specimen.

The patient was admitted to the operating room, under general anesthesia and via the cervical approach, a careful dissection was carried out step by step from the mass to the deep plane which was in direct contact with the jugular axis. carotid, allowing complete resection of the tumor. The postoperative course was simple and uncomplicated. (Fig. 2)

The anatomopathological study of the operative specimen revealed a benign hypocellular multi-nodular tumor proliferation rich in cracked collagen and dilated vessels, with short bundles without particular architecture and infiltrating the peripheral fatty tissue. Tumor cells are spindle-shaped devoid of mitotic activity and atypia, with poorly limited cytoplasm and stroma rich in collagen bundles. On immunohistochemistry, tumor cells express CD 34, but they do not express B catenin, they are negative for smooth muscle actin and desmin. (Fig. 3)

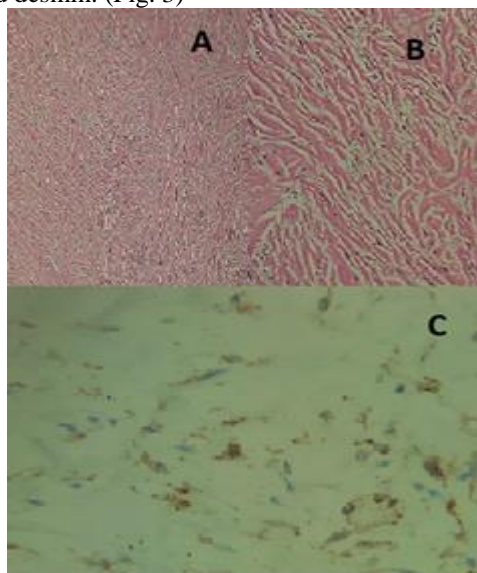


Figure 3. (A) Photomicrograph showing low cell density spindle cell tumor proliferation developing into collagen abundant fibrous stroma forming collagen clusters (HE to GX10). (B) Tumor cells are endowed with regular pycnotic nuclei, without figures of mitosis (HE au Gx20). (C) On immunohistochemistry, cells express CD34.

Given the association between Gardner's fibroma and FAP, we requested upper and lower gastrointestinal endoscopy for recto-colic polyposis with a family investigation. At the end of this assessment, we concluded with the diagnosis of a sporadic Gardner fibroma isolated in front of the absence of family history suggestive of FAP and the absence of mutation of the APC gene.

Discussion

Fibromatous soft tissue lesions, including desmoid-type fibroma and Gardner's fibroma, may occur sporadically or as part of FAP. Those associated with familial adenomatous polyposis often exhibit overexpression of β -catenin. (1,2)

Gardner's fibroma has been defined as a benign soft tissue lesion consisting of randomly arranged bundles of collagen with scattered fibroblasts and plaque-like growth with infiltration of surrounding structures. (3) The sex ratio almost equal to 1 with 78% of cases being diagnosed before the age of 10 years. The most frequent location is the back in a para-vertebral situation. (4,5)

Clinically, appears as a poorly circumscribed subcutaneous fibroadipose mass, rubbery from 1 to 10 cm. In some patients, these fibroids can be multiple and precede the onset of Gardner syndrome by several years. Familial adenomatous polyposis due to a mutation in the APC gene, combines recto-colonic polyposis and several types of benign

tumors such as osteomas, fibroids, epidermoid cysts or desmoid tumors and dental abnormalities. (6,7)

In the context of FAP, the diagnosis is radiologically obvious but requires pathological confirmation which remains to this day the only definite diagnosis. The imaging of choice is MRI. (6,8)

Fibrolipoma, lipomatosis, fibromatosis and elastofibroma are differential diagnoses of this tumor, but the two main ones are nuchal type fibroma and desmoid. Magnetic resonance imaging revealed a tumor that was poorly limited in T1 and T2 hypo signal without enhancement after injection of gadolinium. (9,10)

Histologically, made up of thick collagen bundles randomly arranged between which occasional fibroblasts are found, and having the margins that frequently engulf surrounding structures including fat, muscle, and adjacent nerves. No cytological atypia or necrosis. Immuno histochemically, fibroblasts express beta-catenin and CD34, with a negativity for smooth muscle actin and desmin. (11,12) However, Coffin et al (1) have identified two patients, who had FAP with negative beta-catenin.

Surgery is the basis of treatment, but the very frequent recurrences have led experts to adopt non-surgical therapeutic strategies such as active surveillance. (10)

Pediatric age should alert clinicians to the possibility of underlying Gardner syndrome, hence the interest of an investigation looking for FAP in the patient and his relatives. Regular and close monitoring of the patient is necessary to prevent the risk of developing colonic adenocarcinoma. (10,11)

Conclusion

Gardner's fibroma is a special rare form that can easily be overlooked. Its ignorance can lead to diagnostic delays. There may be the sentinel presentation of Gardner's syndrome and familial adenomatous polyposis that needs to be explored due to the increased risk of colorectal cancer. The basic treatment is surgical, but the evolution remains unpredictable.

Competing interests

The authors declare no competing interests.

References

- Coffin CM, Hornick JL, Zhou H, Fletcher CD: Gardner fibroma: a clinicopathologic and immunohistochemical analysis of 45 patients with 57 fibromas. *Am J Surg Pathol* 2007; 31: 410-416
- Cheryl M. Coffin, Jason L. Hornick, Holly Zhou, and Christopher DM Fletcher, Path.G Gardner Fibroma: A Clinicopathologic and Immunohistochemical Analysis of 45 Patients With 57 Fibromas. *Am J Surg Pathol* Volume 31, Number 3, March 2007
- Essia Saïji, Louis Guillou. Fibroblastic and myofibroblastic tumors of the head and neck. *Annales de pathologie* (2009) 29, 335-346
- Last updated Nov. 6, 2019 gardner fibroma.
- Panjwani S, et al. Gardner's syndrome. *J Clin Imaging Sci* 2011; 1:65
- Bret M. Wehrli, Sharon W. Weiss, Suzanne Yandow, and Cheryl M. Coffin. Gardner-Associated Fibromas (GAF) in Young Patients. *The American Journal of Surgical Pathology* 25 (5): 645-651, 2001
- Joana Vieira, Carla Pinto, Mariana Afons, Maria do Bom Sucesso, Paula Lopes, Manuela Pinheiro, Isabel Veiga, Rui Henrique and Manuel R Teixeira. Identification of previously unrecognized FAP in children with Gardner fibroma. *European Journal of Human Genetics* (2014) /

8. Claudia Santoro, Teresa Giugliano, Delfina Bifano, Carolina D'Anna, Vittoria D'Onofrio Silverio Perrotta. From Gardner fibroma diagnosis to constitutional APC mutation detection: a one-way street. *linical Case Reports* 2017; 5 (10): 1557–1560

9. N. Guignard, C. Cartier, L. Crampette, M. Akkari. Gardner syndrome revealed by a fibromatous tumor of the parotid gland. *French Annals of Otorhinolaryngology and Head and Neck Pathology* 133 (2016) 325–327.

10. Cherif Tayeb Tayeb, Yann Parc 2,3, Thierry Andre, Daniel Lopez-Trabada Ataz. Familial adenomatous polyposis, desmoid tumors and Gardner syndrome. *Bull Cancer* (2019)

11. Rohan Sebastian Pinto, Anthony Simons, Rohit Verma, Neil Bateman. Gardener-associated fibroma: an unusual cause of upper airway obstruction. Pinto RS, et al. *BMJ Case Rep* 2018. doi: 10.1136 / bcr-2018-225079

12. Weiss SW, Goldblum JR. In: Weiss SW, Goldblum JR, editors. *Enzinger and Weiss's Soft tissue tumors*. Fifth edition MosbyElsevier; 2008.