## **Case Report**

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# Adult with colon alveolar rhabdomyosarcoma

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#### **ABSTRACT**

The Purpose of the present study is to present a case of alveolar rhabdomyosarcoma in an 81-year-old patient. An 81-year-old woman presents changes in bowel habits, as well as a mobile mass in the abdomen, fixed to deep planes. Resection is performed. Immunohistochemistry was positive to Myo D1, Anti CD99 and Synaptophysin. Rhabdomyosarcoma may be present in adults, for which it should be discarded in case of small round blue cell tumors. Immunohistochemistry is useful to make a differential diagnosis of the tumor. Survival rates rises when using chemotherapy and surgery.

**Keywords:** Abdominal pain, Alveolar rhabdomyosarcoma, Case report, Rhabdomyosarcoma, Colon cancer, Immunohistochemistry

### INTRODUCTION

This is the case report of an alveolar rhabdomyosarcoma located in the ascendant colon of an 81-year-old patient. Is considered a very rare solid tumor in adults. Rhabdomyosarcoma is the third solid tumor most common in children. Usually (65%) is present in children under 6 years old and in the rest of patients from 10 to 18 years old. In adults it represents less than 3% of solid tumors. Clinically, rhabdomyosarcoma presents a symptomatology according the place it is located, the tumor extension, and the presence or absence of metastasis, for which the diagnosis depends primarily on the histopathology and the tumor markers detected.

### CASE REPORT

The patient is an 81-year-old woman. For significant background, she has in her family Diabetes Mellitus 2 and Systematic Arterial Hypertension. She alludes to

have Irritable Bowel Syndrome of long development, treated with natural supplements (Herbalife®).

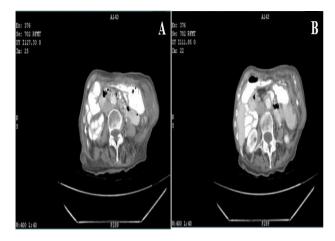


Figure 1: A) TAC contrast fase, B) Irregular reinforcement.

She started her current condition 2 years ago with changes in her bowel habits, with a decrease in the frequency and increase in the consistency, leading to a Bristol 1 once every 3 days.

Eight months previous to diagnosis, she noticed an increase in the hipogastric volume, with a feeling of fullness in the stomach, without pain, denies loss of weight, fever or other discomfort.

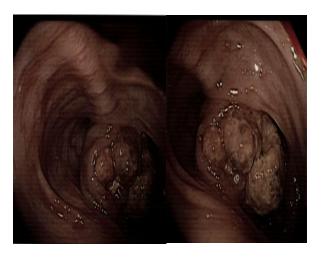


Figure 2: Biopsy used to highlight the tumour with necrotic areas.

When exploring the abdomen, it is soft with normal peristalsis, without tenderness, and there are no changes in color. We palpate a mobile mass in the hypogastrium, approximately of 4 x 4 cm, fixed to deep planes; there are no visceromegalies, no palpable inguinal adenomegalies.

An Abdominal-pelvic Helicoidal Tomography is performed in simple phase and contrast, which reports a collapsed stomach, slightly assessable; concentric mural thickening of the ascending colon and cecum that affects approximately 10 cm, with tightness of the lumen. We also observe irregular reinforcement in the wall with medium of contrast; there are noticeable lymph nodes in the mesenteric fat of 5mm in the short axe, as well as inguinal lymph nodes of 8mm in the short axe (Figure 1A and 1B).

Subsequently, a colonoscopy is performed, finding the rectum with a mucous form and a normal vascular pattern. In the sigmoid colon found multiple diverticula is found, with mucous and a vascular pattern without alterations.

In the colon's splenic angle, there is a tumor with necrotic areas, taking 90% of the lumen, which blocks the pass of the scope. This is why several biopsies are taken and the procedure is concluded (Figure 2).

This first histopathological study was not conclusive, showing only non-specific inflammatory process. This offers the patient open surgery in order to have the resection of the neoplastic mass. The ascending and transversal colon portion is resected, which is sent to the inmuno-histpathological study and the procedure is finalized by making a colon anastomosis.

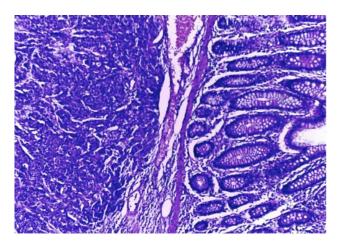


Figure 3: Mucous transversal cut. To the right, the mucous is virtually intact. To the left, solid neoplastic nests, with well-defined borders that infiltrate the sub mucous (HE 40x).

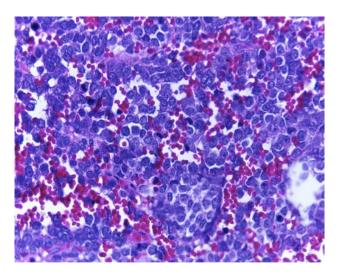


Figure 4: Neoplastic cells with embryonic aspect: small, blue, with little cytoplasm, clear nucleus, reinforced and basophil nucleolus (HE 60x).

The inmuno-histopathological analysis of the pieces sent show neoplastic infiltration in the mucous (Figure 3), as well as an embryonic aspect of the neoplastic cells (Figure 4), with areas of abundant extracellular matrix, which is separated by some underlying fibers, making structures that look like glandular structures with abundant mucous production (Figure 5).

We used inmunoperoxidase Bio and SB staining, which showed positive to Mio D1, Anti CD 99 and Sinaptofisine (Figure 6 and 7); with less intensity for Cytokeratin AE1/AE3 (positive +) and negative to CK20, CD 45, CD 20, CD 3, Chromogranin, MPO, Myogenin,

Protein S100, HMB 45, FLY 1, ALK-1 and CD 30 (Figure 8).

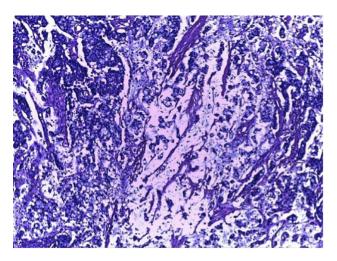


Figure 5: The cells form tiny structures that seem glands (HE 40x).

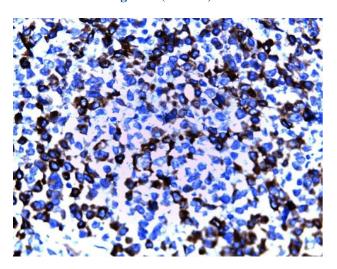


Figure 6: Antibodies against Mio D1, Positive +++, dark ochre coloring.

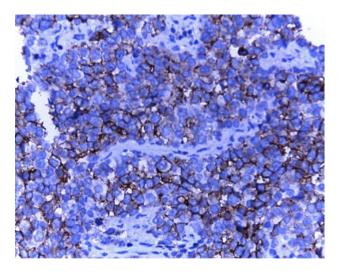


Figure 7: Anti CD 99, positive +++.

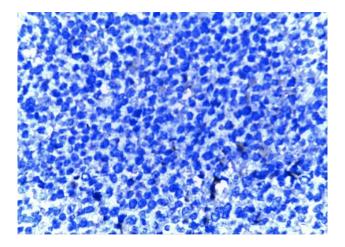


Figure 8: Negative indicators.

An accurate diagnosis of an Alveolar Rhabdomyosarcome in the colon of an 81-year-old woman was established with the information above.

#### DISCUSSION

Rhabdomyosarcoma is the third solid tumor most common in children. Usually (65%) is present in children under 6 years old and in the rest of patients from 10 to 18 years old. In adults it represents less than 3% of solid tumors. 1,2

Rhabdomyosarcoma is a solid tumor derived from the embryonic mesenchyma, which has a differentiation capacity into cells from the musculoskeletal tissue.<sup>2</sup> The rhabdomyosarcoma course is usually of fast growth, with easy hematogenous and lymphatic dissemination, as well as metastasis of regional lymph nodes, heart, lungs and bone. Up to today, there are no known risk factors or predisposition for the appearance of the rhabdomyosarcoma.<sup>4</sup>

In pediatric patients, its appearance is more frequent in head and neck (35-40%), the genitourinary tract (20%) and extremities (15-20%).<sup>5</sup> In adults, the more frequent places have not been described with the same precision; Khosla and cols, describe similar places to the pediatric ones, although the authors mention other places with more frequency.<sup>3,6,7</sup>

Due to its histologic characteristics, it can be grouped in the classification of small round and blue cells, because with the staining hematoxilin-eosin, we observe multiple cells with big dark blue nucleus with reduced cytoplasm.<sup>6</sup> It is necessary to observe through the microscope the striations of the skeletal muscle and an essential part includes visualization of the muscle's own proteins, such as desmin, actin, myoglobin, myosin and MyoD.<sup>3,4</sup>

The rhabdomyosarcoma histopathology classification is done based on cellular differentiation, growth pattern and the extension of cells of embryonic, alveolar and pleomorphic types. Embryonal variety is more common during childhood (almost 70%) and it is sub-classified in botryoids and fusocellular, with preferential location in the head, neck and genitourinary system. Alveolar variety is more frequent in patients from 10 to 18 years old, presented in extremities. This presents fibrous septa that separate cellular groups resembling alveolus. Pleomorphic variety has been found in elderly patients and has a high grade of malignancy. 1,4,5

Immunohistochemistry is a fundamental tool for diagnosis, since, due to the mesenchymal and therefore muscular origin the myogin, desmin and vimentin could be positive in these tumors. However, a differential diagnosis has to be done with other maligned tumors within the group of small round and blue cells tumors (ex. Ewing Sarcoma, lymphomas, neuroblatomas, etc).<sup>4,5</sup>

Clinically, rhabdomyosarcoma presents a symptomatology according the place it is located, the tumor extension, and the presence or absence of metastasis, for which the diagnosis depends primarily on the histopathology and the tumor markers detected.<sup>3</sup>

Nowadays, the rhabdomyosarcoma in a pediatric age is curable in most cases, especially if they receive combined treatments. There is a live expectancy of five years above 70%. In children, botryoids variety presents a better prognosis that the others, being the alveolar the worst prognosis. In adults the alveolar type has a better prognosis than in children.

There have been described some factors that determine a bad prognosis in alveolar sarcoma as it is when presented in adults, an advance stage or its presence in the extremities. However, a medical-surgical treatment with chemotherapy has increased the survival rate up to 5 years in 45% of the cases, and there is evidence that in some cases, the response rate to chemotherapy in adults is similar to pediatric patients. <sup>6,7,9,10</sup>

### **CONCLUSION**

Even though rhabdomyosarcoma is more frequent in patients within pediatric age, it is also present in adults.

However, its appearance does not have a defined pattern, for which it has to be dismissed in the event of small round and blue cells suspicion and having a differential diagnosis with immunohistochemistry techniques. The treatment has to be combined with chemotherapy and

surgery, since this has been proven to increase patients' survival.

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