Faviana et al., J Clin Case Rep 2019, 9:5
DOI: 10.4172/2165-7920.10001238

Journal of Clinical Case Reports

Case Report Open Access

Pleomorphic Rhabdomyosarcoma with Neuroendocrine Differentiation in Abdominal Wall

Faviana P*, Bartolucci A, Boldrini L, Musco B, Farci F, Ferrari M, Ricci A and Lippolis P

Department of Surgical, Medical, Molecular Pathology and Critical Area, University of Pisa, Via Roma, Pisa, Italy

Abstract

The present study presents a case of primary pleomorphic rhabdomyosarcoma that occurs in the abdominal wall. A 52-year-old patient arrived in our department. In his clinical history they have been: 2016 advanced serous ovary papillary carcinoma; a diagnosis of a primary gynecological tumor with secondary extension, and the patient was prepared for bilateral total abdominal hysterectomy salpingo-oophorectomy, omentectomy. Macroscopic and histopathological evaluation of the specimen removed surgically showed a medially differentiated serous papillary carcinoma. The patient received Carbo-Taxol and avastin as postoperative chemotherapy. Postoperative follow-up and CT twelve months after surgery, revealed signs of tumor recurrence: two multilobate neoformations of 15 cm and 5 cm in size respectively with intestinal obstruction. The final histological diagnosis was pleomorphic rhabdomyosarcoma with neuroendocrine differentiation.

Keywords: Ovary cancer; Pleomorphic rhabdomyosarcoma; Immunohistochemistry

Introduction

Rhabdomyosarcomas (RMS) more commonly afflict children and adolescents. It is rare in adults, accounting for 1% of all soft tissue sarcomas. In adults, rhabdomyosarcomas are embryonal (34%), alveolar (23%) or pleomorphic (43%), rarely spindle cell or sclerosing. Adult-type excludes embryonal and alveolar types. Most so-called abdomyosarcomas in adults within the internal trunk are in fact dedifferentiated liposarcomas with heterologous rhabdomyoblastic differentiation. Clinical features: true adult rhabdomyosarcomas occur predominantly in the lower limb, trunk wall or upper limb. RMS has been divided into 3 main subtypes: Embryonal, alveolar and pleomorphic RMS (PRMS). The most common subtypes are the embryonal and alveolar subtypes. Primary PRMS is relatively rare and primarily affects adults, with a peak incidence in the fifth decade of life. It most commonly arises in the deep soft tissues of the extremities. Sarcomas showing neuroendocrine/neuronal differentiation are uncommon: apart from tumors such as peripheral primitive neuroectodermal tumor (pPNET) [1-5] and malignant gastrointestinal stromal/autonomic nerve tumor (plexosarcoma) [6], examples include extra skeletal myxoid chondrosarcoma [7-9] malignant peripheral nerve sheath tumor and ectomesenchymoma [10-12]. This report provides the detailed clinicopathologic findings of 1 case of pleomorphic rhabdomyosarcoma (PRMS) showing neuroendocrine/neuronal differentiation based on classical techniques of histology and immunostaining.

Case Report

In September 2017 a 52-year-old female patient presented to our department. In her clinical history were: 2016 advanced ovary papillary serous carcinoma; a diagnosis of a primary gynecologic tumor with secondary extension, and the patient was prepared for total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy. The macroscopic and histopathological assessment of the surgically resected specimen showed a midly differentiated papillary serous carcinoma, showing enlarged cell nuclei with prominent nucleoli and abundant mitoses. Architecturally the tumor showed growth pattern, with papillary structures. Immunohistochemistry showed positive immunostaining for WT1, CK7, ER and PR. Based on the histomorphology and the immunohistochemical profile of the tumor, pathological assessment concluded that the specimen was a midly differentiated (high-grade) papillary serous carcinoma. For postoperative chemotherapy she received Carbo-Taxolo and avastin.

2017 Postoperative follow-up and CT twelve months after surgery revealed two multilobate mass respectively 15 cm and 5 cm in size with intestinal occlusion. Surgical treatment consisted of right hemicolectomy extended to the transverse medium, paracellular resection of the small intestine, segmental peritonectomy, completion of the perisplenic omentectomy, cholecystectomy, and peritoneal nodules. The resected tumor tissue was fixed in 10% formalin, embedded in paraffin and cut into 5 µm sections using a microtome. The sections were subsequently stained with hematoxylin and eosin and visualized under a microscope. The tumor displayed cellular admixtures of pleomorphic spindle cells and polygonal, rhabdomyoblastic cells arranged in poorly defined clusters. The spindle cells were configured in vague fascicles and formed the background proliferation in which variable numbers of polygonal cells were distributed. The ratio of spindle to polygonal cells was at least 10:1, but the latter cells were easily identifiable in almost every high-power microscopic field. The polygonal cells displayed abundant, brightly eosinophilic cytoplasm and eccentrically located, pleomorphic nuclei. Both cellular populations displayed marked cytologic atypia, with marked anisonucleosis, abnormalities of chromatin, and nuclear membrane irregularities. Osteoclast-like giant cells (Figure 1) and numerous mitotic figures were present, including highly atypical forms (average mitotic index: 30-40 mitotic figures per 10 high power fields). The cells revealed scanty to moderate amounts [13] of pale eosinophilic cytoplasm with indistinct borders. The nuclei were vesicular, moderately large, round to by Oliveira et al. [7] oval with clumped or coarse chromatin and contained one or more prominent nucleoli. Microscopic analysis showed full-thickness infiltration of the colon and small intestine wall and diffuse infiltration of the peritoneum.

***Corresponding author:** Faviana P, Department of Surgical, Medical, Molecular Pathology and Critical Area, University of Pisa, Via Roma, Pisa, Italy, Tel: +39 3337830811; E-mail: pinuccia.faviana@med.unipi.it

Received April 16, 2019; **Accepted** April 24, 2019; **Published** May 01, 2019

Citation: Faviana P, Bartolucci A, Boldrini L, Musco B, Farci F, et al. (2019) Pleomorphic Rhabdomyosarcoma with Neuroendocrine Differentiation in Abdominal Wall. J Clin Case Rep 9: 1238. doi: 10.4172/2165-7920.10001238

Results

Immunohistochemical reactions for cytokeratins (AE1/3, CK7, CK20, CAM5.2), WT-1, CA125, MART-1, S100 all found negative. In contrast, vimentin, desmin, actin, myogenin (Figure 2), reacted with tumor cells that show a diffuse neuroendocrine differentiation: synaptophysin, NSE and cromogranin (Figure 3) positive. The tumor was negative for estrogen and progesterone. The proliferation index was very high, revealing 30-40 mitoses for 10 high power fields, that was demonstrated by immunostaining with anti-Ki-67 antibodies. The final diagnosis was pleomorphic rhabdomyosarcoma with neuroendocrine differentiation. The the patient died about 90 days after the first hospitalization due to organ failure. For postoperative chemotherapy he received only avastin, non-reactive and with rapidity progression to death approximately 2 months after surgery.

Discussion

RMS is divided into 3 main subtypes: Embryonal and alveolar RMS and PRMS, according to the 2002 World Health Organization Classification of Soft Tissue and Bone Neoplasms [14-21]. PRMS was first described by Stout in 1946. Primary PRMS is relatively rare and primarily affects adults in the fifth decade of life and are mostly located in the lower extremity. Less frequent sites of presentation are the abdomen, retroperitoneum, chest wall, spermatic cord/testes, upper extremity, mouth, and orbit. They typically have an aggressive clinical course, demonstrating an overall poor prognosis [22,23]. The histological manifestations of RMS widely vary, and the histopathological diagnosis is based on morphological and immunohistochemical stains that reveal a skeletal muscle phenotype. PRMS Morphologically, were composed of large, atypical, polygonal pleomorphic rhabdomyoblasts with abundant eosinophilic cytoplasm. These large rhabdomyoblasts are often arranged in clusters, sheets, or scattered individual cells. Atypical, vesicular nuclei with prominent nucleoli predominate. The rhabdomyoblasts in the background that surround the large, pleomorphic rhabdomyoblasts vary from round to spindled. Neuroendocrine/neuronal differentiation was not [7] demonstrated completely in rhabdomyosarcoma. This study wants to highlight [7] the neuroendocrine/neuronal differentiation in rhabdomyosarcoma, and specifically the pleomorphic variant [24-26].

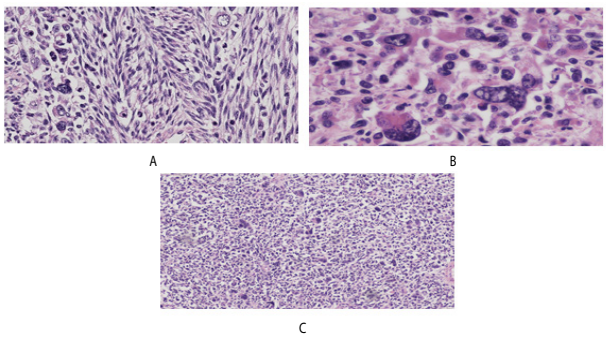

Figure 1: (A) The tumor displayed cellular admixtures of pleomorphic spindle cells and polygonal, rhabdomyolysis cells arranged in poorly defined clusters, (B) polygonal cells with abundant, brightly eosinophilic cytoplasm and eccentrically located, pleomorphic nuclei, (C) osteoclast-like giant cells were present.

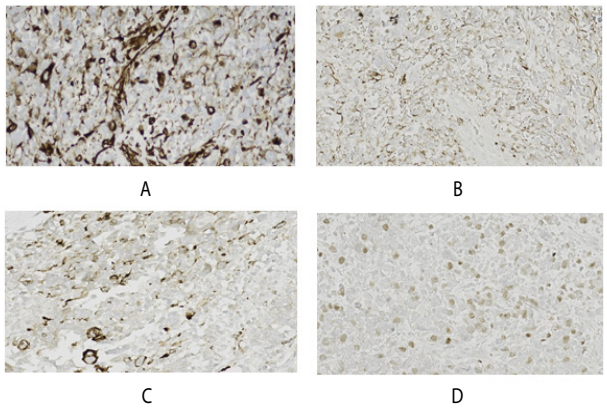

Figure 2: The myogenic phenotype is highlighted by immunohistochemical stains for (A) vimentin, (B) desmin, (C) actin HHF35, (D) myogenin.

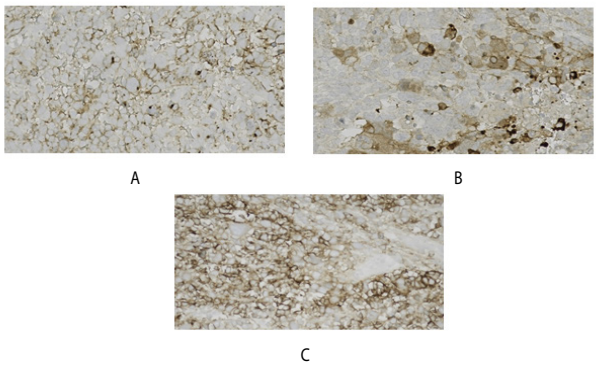

Figure 3: The neuroendocrine phenotype is highlighted by immunohistochemical stains, tumor cells demonstrate strong positivity for synaptophysin (A), NSE (B) and chromogranin (C).

Conclusion

The positive immunoreactivity for myogenic markers supported rhabdomyoblastic differentiation. According to all classical criteria, therefore, the tumor conform to PRMS. In the definition of neuroendocrine differentiation in tumors, in addition to the histologic features, chromogranin and synaptophysin immunostaining forms the most practical and widely used criterion. Chromogranin and synaptophysin are widely regarded as reliable and specific. In rhabdomyosarcoma, neurone specific enolase (NSE), and CD56 have been demonstrated; these markers were originally thought to be specific for neuroendocrine differentiation, but their specificity has been brought into question by their demonstration in a wide variety of non neuroendocrine cells. These papers show carcinomas with neuroendocrine and rhabdomyosarcomatous differentiation. To our knowledge this is a rare case report of abdomen wall pleomorphic rhabdomyosarcoma. The presentation of a rare adult sarcoma with neuroendocrine differentiation mimicking a gynecologic malignancy was an unusual feature that complicated the diagnosis in this case.

References

1. Pastore G, Peris-Bonet R, Carli M, Martinez-Garcia C, De-Toledo SJ, et al. (2006) Childhood soft tissue sarcomas incidence and survival in european children (1978-1997): Report from the automated childhood cancer information system project. Eur J Cancer 42: 2136-2149.

2. Borka K, Patai K, Rendek A, Sobel G, Paulin F (2006) Pleomorphic rhabdomyosarcoma of the uterus in a postmenopausal patient. Pathol Oncol Res 12: 102-104.

3. Fletcher CD, Bridge JA, Hogendoorn P, Mertens F (2013) WHO classification of tumours of soft tissue and bone (IARC WHO classification of tumours). World Health Organisation 2: 1.

4. Enzinger FM (2001) Primitive neuroectodermal tumors and related lesions. Soft Tissue Tumors pp: 1289-1321.

5. Pagani A, Macri L, Rosolen A (1998) Neuroendocrine differentiation in Ewing's sarcomas and primitive neuroectodermal tumors revealed by reverse transcriptasepolymerase chain reaction of chromogranin mRNA. Diagn Mol Pathol 7: 36-43.

6. Herrera GA, Cerezo L, Jones JE (1989) Gastrointestinal autonomic nerve tumors: Plexosarcomas. Arch Pathol Lab Med 113: 846-853.

7. Oliveira AM, Sebo TJ, McGrory JE (2000) Extraskeletal myxoid chondrosarcoma: A clinicopathologic, immunohistochemical, and ploidy analysis of 23 cases. Mod Pathol 13: 900-908.

8. Harris M, Coyne J, Tariq M (2000) Extraskeletal myxoid chondrosarcoma with neuroendocrine differentiation: A pathologic, cytogenetic, and molecular study of a case with a novel translocation. Am J Surg Pathol 24: 1020-1026.

9. Goh YW, Spagnolo DV, Platten M (2001) Extraskeletal myxoid chondrosarcoma: A light microscopic, immunohistochemical, ultrastructural, and immuno-ultrastructural study indicating neuroendocrine differentiation. Histopathol 39: 514-524.

10. Huang L, Espinoza C, Welsh R (2003) Malignant peripheral nerve sheath tumor with divergent differentiation. Arch Pathol Lab Med 127: 147-150.

11. Karcioglu Z, Someren A, Mathes SJ (1977) Ectomesenchymoma: A malignant tumor of migratory neural crest (ectomesenchyme) remnants showing ganglionic, schwannian, melanocytic, and rhabdomyoblastic differentiation. Cancer 39: 2486-2496.

12. Kawamoto EH, Weidner N, Agostini RM (1987) Malignant ectomesenchymoma of soft tissue report of 2 cases and review of the literature. Cancer 59: 1791-1802.

13. Kosem M, Ibiloglu I, Bakan V (2004) Ectomesenchymoma: Case report and review of the literature. Turk J Pediatr 46: 82-87.

14. Horn RC, Enterline HT (1958) Rhabdomyosarcoma: A clinicopathological study and classification of 39 cases. Cancer 11: 181-197.

15. Albores-Saavedra J, Martin RG, Smith JL (1963) Rhabdomyosarcoma: A study of 35 cases. Ann Surg 157: 186-197.

16. Stout AP (1946) Rhabdomyosarcoma of skeletal muscles. Ann Surg 123: 447-472.

17. Ariel IM, Briceno M (1975) Rhabdomyosarcoma of the extremities and trunk: Analysis of 150 patients treated by surgical resection. J Surg Oncol 7: 269-287.

18. Keyhani A, Booher RJ (1968) Pleomorphic rhabdomyosarcoma. Cancer 22: 956-967.

19. Linsheid RL, Soule EH, Henderson ED (1965) Pleomorphic rhabdomyosarcoma of the extremities and limb girdles: A clinicopathologic study. J Bone Joint Surg 47: 715-726.

20. Pack GT, Eberhart WF (1952) Rhabdomyosarcoma of skeletal muscle: Report of 100 cases. Surg 32: 1023-1064.

21. Patton RB, Horn RC (1962) Rhabdomyosarcoma: Clinical and pathological features and comparison with human fetal and embryonal skeletal muscle. Surg 52: 572-584.

22. Phelan JT, Juardo J (1962) Rhabdomyosarcomas. Surg 52: 585-591.

23. Furlong MA, Mentzel T, Fanburg-Smith JC (2001) Pleomorphic rhabdomyosarcoma in adults: A clinicopathologic study of 38 cases with emphasis on morphologic variants and recent skeletal musclespecific markers. Mod Pathol 4: 595-603.

24. Gaffney EF, Dervan PA, Fletcher CD (1993) Pleomorphic rhabdomyosarcoma in adulthood. Analysis of 11 cases with definition of diagnostic criteria. Am J Surg Pathol 17: 601-609.

25. Mechtersheimer G, Staudter M, Moller P (1991) Expression of the natural killer cell-associated antigens CD56 and CD57 in human neural and striated muscle cells and in their tumors. Cancer Res 51: 1300-1307.

26. Leader M, Collins M, Patel J, Henry K (1986) Antineuron specific enolase staining reactions in sarcomas and carcinomas: Its lack of neuroendocrine specificity. J Clin Pathol 39: 1186-1192.