ACG CASE REPORTS JOURNAL

IMAGE | SMALL BOWEL

Adult Duodenal Pleomorphic Rhabdomyosarcoma

Yuto Shimamura, MD[1], Fumio Omata, MD, MPH, PhD[2,3], Kenji Nakamura, MD[1], Seiji Ohigashi, MD, PhD[4], Koyu Suzuki, MD[5], and Yoshiyuki Fujita, MD[1]

[1]*Division of Gastroenterology, St. Luke's International Hospital, Tokyo, Japan*
[2]*Division of Endoscopy, St. Luke's International Hospital, Tokyo, Japan*
[3]*Center for Clinical Epidemiology, St. Luke's Life Science Institute, Tokyo, Japan*
[4]*Division of Surgery, St. Luke's International Hospital, Tokyo, Japan*
[5]*Division of Pathology, St. Luke's International Hospital, Tokyo, Japan*

Case Report

A 73-year-old woman was admitted with severe anemia. An esophagogastroduodenoscopy revealed the presence of a tumor in the second portion of the duodenum (Figure 1); biopsies showed pleomorphic rhabdomyosarcoma (RMS; Figure 2). Phosphotungstic acid-hematoxylin staining highlighted cytoplasmic cross in tumor cells, and immunohistochemical staining demonstrated that the tumor was positive for muscle-specific (desmin) and rhabdomyogenic markers (myoD1 and myogenin), which provided evidence supporting myogenic differentiation (Figure 3). The patient underwent subtotal stomach-preserving pancreaticoduodenectomy without any complications. There has been no tumor recurrence for at least 6 months after surgery.

Identification with light microscopy of rhabdomyoblasts or cross striations characteristic of skeletal muscle is required for the diagnosis of RMS. Since it is often difficult to diagnose RMS, advanced immunohistochemical examination may be needed for definitive diagnosis.[1] The World Health Organization classifies RMS into 4 major histologic subtypes: embryonal, alveolar, pleomorphic, and spindle cell/sclerosing rhabdomyosarcoma.[2] Histologic subtyping is important, as prognosis and clinical behavior can vary by subtype.[3] Pleomorphic RMS is more common in adults, and typically has a poor prognosis.[3] This is the first report of pleomorphic RMS confirmed by positive immunohistochemical staining with myoD1 and myogenin, which are nuclear regulatory proteins specific for skeletal muscle differentiation.

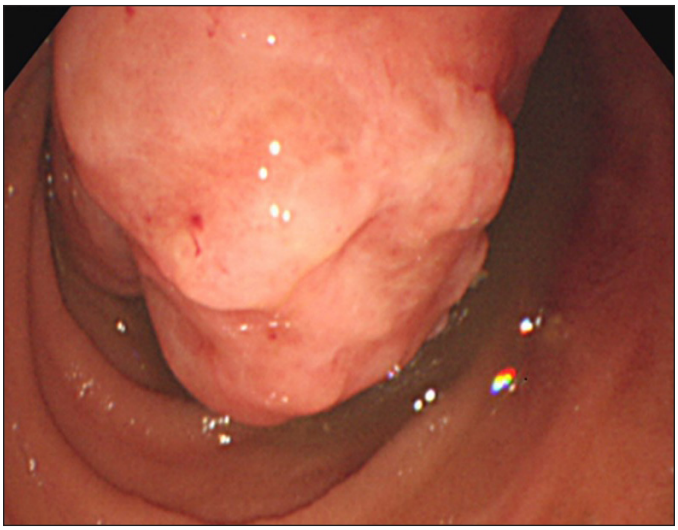

Figure 1. Esophagogastroduodenoscopy showing a protruding lesion close to the major papilla, approximately 50 mm in diameter.

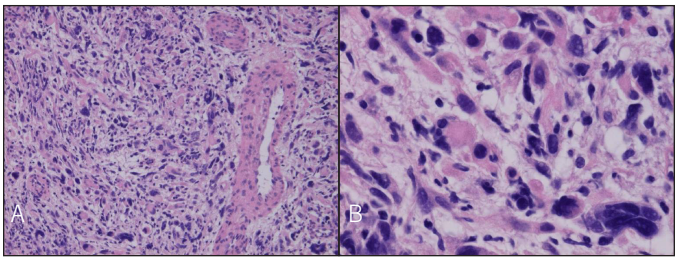

Figure 2. Anaplastic malignant neoplasm composed of proliferating atypical spindle cells with pleomorphic and hyperchromatic nuclei and eosinophilic cytoplasm, with scattered giant multinucleated tumor cells. (A) H&E stain, x10 magnification and (B) H&E stain, x40 magnification.

Disclosures

Author contributions: Y. Shimamura performed the literature review and wrote the initial manuscript. F. Omata critically revised the manuscript and is the article guarantor. Y. Shimamura, K. Nakamura, S. Ohigashi, K. Suzuki, and Y. Fujita

ACG Case Rep J 2014;2(1):11–12. doi:10.14309/crj.2014.67. Published online: October 10, 2014.

Correspondence: Yuto Shimamura, Division of Gastroenterology, St. Luke's International Hospital, 9-1 Akashi-cho, Chuo-ku, Tokyo 104-8560, Japan (yutshi@luke.ac.jp).

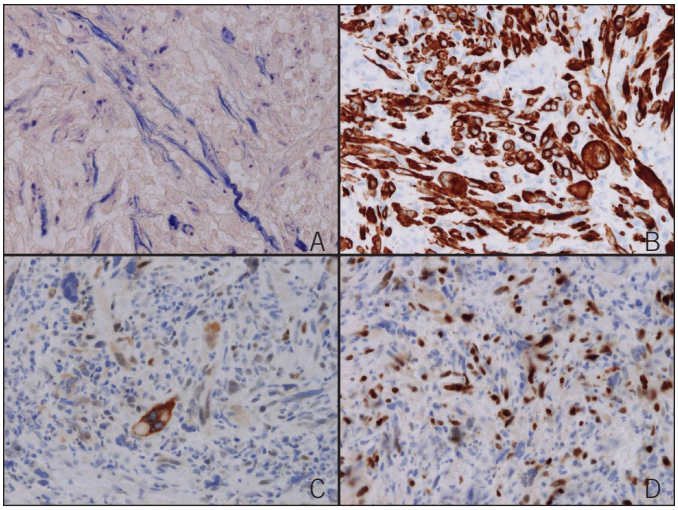

Figure 3. (A) Phosphotungstic acid-hematoxylin stain highlighting the cross striations of the cytoplasm in the tumor cells; (B) desmin, a muscle-specific marker, is positive in the tumor cells; (C) myoD1, a rhabdomyogenic marker, is positive in the tumor cells; (D) myogenin, a rhabdomyogenic marker, is positive in the tumor cells.

provided clinical care and guidance for drafting manuscript.

Financial disclosure: None to report.

Informed consent was obtained for this case report.

Received: July 17, 2014; Accepted: August 14, 2014

References

1. Parham DM, Ellison DA. Rhabdomyosarcoma in adults and children: An update. *Arch Pathol Lab Med*. 2006;130(10):1454–65.
2. Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F, eds. *World Health Organization Classification of Tumours of Soft Tissue and Bone*. 4th ed. Lyon, France: IARC Press; 2013.
3. Furlong M, Mentzel T, Fanburg-Smith JC. Pleomorphic rhabdomyosarcoma in adults: A clinicopathologic study of 38 cases with emphasis on morphologic variants and recent skeletal muscle-specific markers. *Mod Pathol*. 2001;14(6):595–603.