TITLE

Giant desmoid fibromatosis of pancreas

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Short title: Desmoid tumor

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Patient consent: obtained

Author contributions: J-C.C treated the patient, wrote the manuscript and constructed the figures. S-C. H took the photographs of surgical specimens and tumor histology.

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A 15-year-old girl had abdominal distension and appetite loss with episodic heartburn and vomiting for approximately 2 months, but gained 3 kg of weight. Physical examination revealed a huge and hard abdominal mass. Tumor markers including α-fetoprotein, β-human chorionic gonadotropin, carcinoembryonic antigen, CA125, and CA19-9 were all normal. Computed tomography disclosed a gigantic solid mass, occupying the whole abdominopelvic cavity (Figure 1). Notably, the pancreas was only identifiable by its head and contiguous splenic vein. However, this tumor neither involved other intraabdominal vital structures nor caused paraaortic lymphadenopathy. Core needle biopsy showed hypocellular collagenized spindle-cell neoplasm without nuclear hyperchromasia and atypia, suggestive of desmoid fibromatosis (DF). This tumor was extirpated surgically (Figure 2A), measuring 36 × 33 × 17 cm and weighing 13.4 kg. Histologically (Figure 2B–D), it infiltrated the normal pancreas and expressed nuclear β-catenin. Postoperatively, colonoscopy found no polyps. So far, she has been tumor-free for 12 months.

DF is characterized by its clonally fibroblastic proliferation and locally infiltrative and aggressive growth, leading to a tendency of local recurrence following resection despite its inability to metastasize. Thus, it has a variable and often unpredictable clinical course. Intraabdominal DF was relatively rare but frequently occurred in familial adenomatous polyposis. Patients tended to be asymptomatic until signs of DF mass effects or local invasion. Generally, active surveillance is the initial approach to DF. However, surgery remains the first-line treatment in the case of the limited surgical morbidities anticipated for symptomatic DF that grows rapidly, as with this patient whose DF expanded on such an unprecedented scale but was considered operable based on radiographic studies. The DF therapeutic algorithm makes no distinction between adults and children.
**Figure 1.** Computed tomography (CT) of the abdomen, axial (left panels), sagittal (middle panels), and coronal (right panels) views, disclosed an exaggeratedly huge solid tumor, which had comparable CT density to the pancreas and kidneys, and slight contrast enhancement (left panels). The pancreas had missing body and tail, leaving the identifiable head (asterisk) and splenic vein (solid arrow). There was neither paraaortic lymphadenopathy nor radiographic evidence of tumor abutment or encasement of major vessels, including superior mesenteric vessels (open arrowheads), inferior vena cava (v), aorta (a), celiac trunk (open arrow), and hepatic portal vessels (solid arrowhead).

**Figure 2.** (A) Cut surface of the resected DF showed a glistening trabecular appearance, and appeared whitish gray. There was no hemorrhage and necrosis. (B) Hematoxylin and eosin staining of specimen sections disclosed hypocellular neoplasm along with normal pancreatic tissues (open arrowheads) in abundant collagenous stroma. (C) In a magnified view, spindle-shaped tumor cells, arranged in well-defined fascicles, exhibited bland nuclear morphology as evidenced by the absence of nuclear hyperchromasia and atypia. (D) Immunohistochemical staining demonstrated the nuclear localization of β-catenin (open arrows) in spindle-shaped tumor cells. The nuclear counterstaining also showed fine chromatin and inconspicuous nucleoli of the tumor cell nuclei.

**Reference**


AUTHORSHIP & CONFLICTS OF INTEREST STATEMENT

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AUTHORSHIP

All persons who meet authorship criteria are listed as authors, and all authors certify that they have participated sufficiently in the work to take public responsibility for the content, including participation in the concept, design, analysis, writing, or revision of the manuscript. Furthermore, each author certifies that this material or similar material has not been and will not be submitted to or published in any other publication.

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acquisition of data (typed): Jeng-Chang Chen, Shih-Chiang Huang;

analysis and/or interpretation of data (typed): Jeng-Chang Chen.

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