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Case study

Giant intra-abdominal mature cystic teratoma (dermoid cyst) in an adult man, with male genitourinary tissue including prostatic and penile elements☆

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Summary We describe a case of a giant intra-abdominal mature cystic teratoma in a 36-year-old man, which comprised typical features of differentiated teratoma/dermoid cyst but which contained a macroscopic rudimentary penis, with vasoformative erectile tissue–like structures consistent with corpora cavernosa, as well as scrotal-type skin and prostatic tissue. The genitourinary structures were well formed both grossly and microscopically and sharply demarcated from the rest of the neoplasm, which comprised typical differentiated teratoma, without any other macroscopic foci of organoid differentiation or of other histologic differentiation. The plasticity of the cells of differentiated teratoma, which enables it to undergo multidirectional differentiation, is well recognized, but the factors determining this distinct path of differentiation remain to be established.

1. Introduction

Teratomas are neoplasms with cell types representative of more than 1 germ cell layer. Mature teratoma is a benign tumor that is usually cystic and commonly contains structures derived from endoderm, ectoderm, and mesoderm [1]. Dermoid cysts (mature cystic teratomas) are a form of mature teratoma of predominantly ectodermal derivation and, by definition, form skin-like structures with adnexal elements, although they may also contain other tissue types [2]. Characteristically, they are unilocular and, in addition to cutaneous-type components, may contain components of other tissues, including hair and teeth, glandular elements, adipose tissue, cartilage, and bone [2], as well as sebaceous material. These are common within the ovary, occurring most frequently in women of reproductive age where they are usually benign, but dermoid cyst–like mature teratomas are rare in males. These neoplasms can also arise in several other sites, including rarely the abdominal cavity and retroperitoneum.

We describe the case of an adult man with an unusual type of giant retroperitoneal dermoid cyst/mature cystic teratoma showing otherwise unremarkable features other than the harboring of several types of male genitourinary tissues constituting an isolated rudimentary penis. The lack of other

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lines of differentiation distinguishes this case from more common differentiated teratomas with multidirectional differentiation, and this is compounded by the fact that ovarian-type dermoid cysts/mature teratomas (including those occurring intra-abdominally) are rare in males. This highly organized pattern of divergent differentiation is a previously undocumented finding, which expands the spectrum of differentiation seen within intra-abdominal mature cystic teratomas, and we discuss the clinical and pathologic findings and the differential diagnosis.

2. Case history

A 36-year-old white man presented acutely with an episode of abdominal pain with hematemesis and melena. He had a longstanding history of gradual increase in his abdominal girth since childhood but had been otherwise asymptomatic, with no symptoms of bowel obstruction or weight loss. He was well with no previous medical or family history. Clinical examination showed a large, nontender abdominal mass but was otherwise unremarkable. He had normal male external genitalia, and both testes were present within the scrotum. Esophagogastroduodenoscopy showed duodenal ulceration at D2, and he was treated with epinephrine injection. Computed tomography scan showed a 32-cm-diameter, partly fatty density and partly cystic thin-walled mass arising centrally from the abdomen and retroperitoneum, which had the radiologic appearance of a dermoid cyst or benign teratoma. At laparotomy, this was a huge, smooth-surfaced fibrous-walled cyst, with stomach and duodenum splayed over its surface. Because of its size, it was not possible to access the abdominal cavity to dissect the bowel from the lesion, so the cyst was opened, and 20 L of contents, comprising fluid and necrotic debris, was manually cleared. The cyst was mobilized from the retroperitoneum but was impossible to separate from medial structures (common bile duct, undersurface of pancreas, and posterior surfaces of stomach and duodenum), so cholecystectomy was performed with hepaticojejunostomy and Roux-en-Y reconstruction. The patient made an uneventful postoperative recovery and is well 14 months after surgery. He has been followed up with serum biochemistry including germ cell markers and prostate-specific antigen (PSA), which remain within the reference range.

3. Materials and methods

The histopathological features were noted, and immunohistochemistry was performed on formalin-fixed, paraffin-embedded (FFPE) material using the following commercial antibodies: CK7, CK20, 34βE12, smooth muscle actin (Dako, Glostrop, Denmark; 1:50), CD31 (DAKO; 1:20), PSA (DAKO; 1:25), prostate-specific acid phosphatase (PSAP) (DAKO; 1:1500), α-methylacyl coenzyme A racemase (Leica Microsystems, Milton Keynes, UK; 1:50), CD34 (Novocastra, Newcastle-upon-Tyne, UK; 1:30), ERG, p63, and GATA3 (Ventana Medical Systems, Inc, [Roche], Tucson, AZ; prediluted). Fluorescence in situ hybridization was performed on FFPE material to assess for evidence of isochromosome 12p. One-micrometer-thick FFPE sections were dewaxed overnight at 60°C, treated with hot buffer wash at 80°C (2-3 hours) and then proteolytic enzyme treatment at 37°C, and washed in distilled water and then an alcohol series before addition of *ETV6*/*RUNX1* DNA single-fusion probes (Vysis, Abbott Laboratories Ltd, Maidenhead, UK). Hybridization was performed overnight according to manufacturer's protocols.

4. Results

4.1. Pathologic findings

Gross examination of the specimen showed a large, collapsed, opened bilocular cyst measuring at least 24 cm (with wall ranging from 0.4 to 1.7 cm in thickness) (Fig. 1A), with foci of fibrinopurulent exudate on the outer surface. A thick intracystic septum was present. The internal surface showed prominent, cobblestone-like congested reddish-purple nodularity. No necrosis was present. Attached to the intracystic septum was a 7.5 × 2.5 × 2.5–cm cylindrical, pale tan structure with a soft, wrinkled surface (Fig. 1A-D). The base of the structure was attached to the cystic septum, with the rest of it standing freely. This structure, for the large part, was covered by rugose, unremarkable skin. There was prominent hair at its base, and at its distal aspect, the surface was coarser, with some roughened, spiky processes. A 0.5-cm pearly nodule was also present near the tip. Other than this cylindrical structure, the rest of the cyst was uniform, with no other solid or papillary areas or hair present. Histologically, this was a bilocular cystic neoplasm with a variably thickened wall. The inner cystic lining comprised keratinizing, stratified squamous epithelium with a granular layer. The epithelium was multifocally ulcerated in an alternating pattern of ulceration and intact epithelium, corresponding with the cobblestone-like appearance seen macroscopically (Fig. 2A). The intact epithelium showed prominent underlying dilation of thin-walled vessels. The ulcerated areas had fibrinopurulent exudate, with underlying granulation tissue and fibrosis.

The cylindrical structure described macroscopically was a shaft-like structure, with its base attached to the inner cyst wall. It was lined by mature keratinizing squamous epithelium with a prominent rugose appearance (Fig. 2B and C). The epithelium near the tip of the structure was hyperkeratotic, papillomatous, and spiky (Fig. 2B). Within the rest of the structure, the subepithelial tissue, corresponding to dermis, contained prominent skin appendageal structures, including eccrine and apocrine glands and pilosebaceous units with hair shafts, smooth muscle bundles, and fat, and the features were highly reminiscent of scrotal skin (Fig. 2C). The pearly nodule