Case report

INCIDENTAL, LOW-FAT VARIANT OF SPINDLE CELL LIPOMA: A NOVEL TUMOUR OF THE SMALL INTESTINE

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A woman underwent surgical intervention for a carcinoma of the ovary. In the intervention, a submucosal nodule of the ileum was found. Pathological study revealed a spindle cell lipoma (SCL). This case revealed the presence of CD34-positive spindle and stellate cells with dendritic cytoplasmic prolongations, a feature shared with dendritic fibromyxolipoma. Fluorescence in situ hybridisation analysis showed 13q14 heterozygous deletion. Spindle cell lipoma of the small intestine has not been previously reported. Spindle cell lipoma, although rare, should be included among the benign mesenchymal lesions of the small intestine. This report extends the range of locations in which this tumour is found to arise.

Key words: small intestine, gastrointestinal tract, spindle cell lipoma, dendritic fibromyxolipoma, immunohistochemistry.

Introduction

Conventional or ordinary lipoma is the most common mesenchymal tumour in adults. However, small intestinal lipomas are uncommon, and their clinical presentation is nonspecific. Thus, in a study of 50 patients with small bowel tumours, only two cases of conventional lipoma (4%) were observed [1]. On the other hand, spindle cell lipoma (SCL) is an uncommon and distinctive variant of lipoma, characteristically arising in the hypodermis of the neck, shoulder, or back of men between 46 and 81 years of age [2]. As far as we are aware, no case of SCL has been reported in the small intestine. In this report, we present a case of this condition.

Clinical history

A 72-year-old woman underwent surgical intervention for the removal of a high-grade serous carcinoma of the left ovary with peritoneal dissemination. In the surgical procedure, a small submucosal nodule of the terminal ileum was found and removed for pathological study. That lesion had not been detected in either the abdominopelvic echography or the abdominopelvic CT scan performed on the patient.

Pathological analysis

The resected specimen was routinely fixed with 45 formalin and processed for paraffin embedding. Sections 4 µm thick were stained with haematoxylin-eosin. Immunopathological studies were carried out on formalin-fixed, 4-µm-thick, paraffin-embedded tissue sections using the EnVision FLEX+ Visualisation System (Dako, Agilent Technologies, SL, Las Rozas, Madrid, Spain). Immunohistochemical reaction was performed using appropriate tissue controls for the antibodies utilised. Automatic staining was performed on an Autostainer Link 48 (Dako, Agilent Technologies, SL). Antibodies used in the immunohistochemical study are detailed in Table I. Fluorescence in situ hybridisation (FISH) analysis was performed on four-microm-thick, paraffin-em-
bedded tissue sections. We used the locus-specific probe Vysis LSI D13S319 SpectrumOrange specific for Rb locus with the control of locus-specific 13q34 SpectrumAcqua (Abbott Molecular, Vysis, Des Plaines, IL, USA) according to the manufacturer’s instructions. In brief, normal cells must present two copies of each probe. In cells where an Rb deletion is present, a two acqua-one orange signal must be detected. Fifty interphase nuclei were evaluated in tumour cells.

Results

The segmental excision of the ileum showed a submucosal nodulation of 14 × 13 × 13 mm, whitish in colour, and firm in consistency. Microscopically the tumour was well circumscribed (Fig. 1A), hypocellular, and with collagenous stroma (Fig. 1B). It was composed of a proliferation of bland spindle or stellate cells with small, uniform, fusiform, or triangulate hyperchromatic nuclei with inconspicuous nucleoli and pale, poorly defined cytoplasm set in a collagenised stroma (Fig. 1C). These cells were predominant and showed a random dispersed arrangement. Isolated, scattered, single mature adipocytes were very scarce (Fig. 1D). The adipocyte count in 40 fields produced a score of 8.3 adipocytes per 10 high-power fields (40× objective), comprising <5% of the lesion. Lipoblasts were not found. Spindle or stellate tumour cells were associated with hyalinated thick rope-like collagen bundles (Fig. 2A). Entrapment of a group of ganglionic cells of submucosal (Meissner’s) plexus was seen (Fig. 2B). Cellular atypia, mitoses, myxoid stroma, and small plexiform vascular proliferation were not observed. There were occasional dispersed mast cells.

Immunohistochemically, the spindle and stellate cells were strongly positive for vimentin (Fig. 2C) and CD34 (Fig. 2D), and negative for S100 protein, epithelial membrane antigen, smooth muscle actin, desmin, BCL2 oncoprotein, neurofilament protein, CD117, DOG-1, GluT1, claudin-1, and STAT6. Numerous vimentin+ and CD34+ cells showed prominent slender, dendritic cytoplasmic prolongations. Mature adipocytes were reactive for S-100 protein. Ki67 staining showed very scant labelling of the tumour. FISH analysis showed 13q14 heterozygous deletion (monoallelic deletion) in a considerable number of counted nuclei (Fig. 3).

Discussion

Gastrointestinal conventional lipomas are mesenchymal, benign, slow-growing, usually single tumours consisting entirely of mature adipocytes. The incidence of lipomas varies depending on whether the study includes autopsy or surgical material. In a collected series of 113,932 autopsies from the literature, 232 cases of gastrointestinal lipoma were observed; an incidence of 0.20% [3]. The most common location is the colon, but they can also be found in the oesophagus, stomach, and small intestine [4]. The most frequent site in the small intestine is the ileum (54%), followed by the duodenum (32%) and jejunum (14%) [5]. Small intestinal lipomas are more frequent in men (68.2%), and the peak age of occurrence is the seventh decade [5]. In a predominantly surgical series, they comprised 26.2% of benign

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EMA – epithelial membrane antigen; SMA – smooth muscle actin; RTU – ready-to-use Dako, Agilent Technologies, SL, Las Rozas, Madrid, Spain; Novocastra, Leica Microsistemas, SLU, Barcelona, Spain; abcam plc, Cambridge, UK; GeneTex Inc., Bellaterra, Barcelona, Spain;

Table I. Antibodies used in this study
small bowel tumours [5]. In general, when these tumours are larger than 2 cm they tend to produce symptoms such as gastrointestinal bleeding, anaemia, intussusception, or bowel obstruction [5, 6].

Computed tomographic (CT) scan and magnetic resonance imaging (MRI) depict lipomas as homogeneous, non-enhancing, well-delimited lesions consistent with adipose tissue [7].

To our knowledge, SCL has not been reported in the small intestine. A unique case of SCL described in the gastrointestinal tract was reported by Robb and Jones in a 65-year-old man. The neoplasm, measuring 4 cm, was located in the perianal region with involvement of the external anal sphincter [8]. Spindle cell lipoma is a lipoma variant variably composed of mature fat, bland, non-lipogenic, CD34-positive spindle cells and coarse, rope-like collagen bands situated between the cellular elements. Spindle cells are also reactive for vimentin and Bcl2. The positivity for Bcl2 is observed in about 56% of cases [9]. Some tumours are fat-dominated with scant spindle cells while others show a prominent spindle cell component with few, or no fat cells. Thus, mature fat cells may be present in < 5% of the tumour (low-fat variant) or even absent (fat-free variant) [10]. These two variants pose a substantial challenge to pathologists.

Fig. 1. Histopathology of the spindle cell lipoma of the small intestine. A) Panoramic view of the entire tumour. Note the circumscription of the lesion. The neoplasm is located in the submucosa of the small intestine. The solitary mature adipocytes are scarce and irregularly distributed (HE, original magnification 12,5×). B) The submucosal neoplasm is hypocellular with collagenised stroma (HE, original magnification 40×). C) Spindle cells are predominant. Occasional mature adipocytes are present (HE, original magnification 100×). D) Spindle cells show a random dispersed arrangement. Numerous ropey collagen bundles are evident. A mature adipocyte can be observed. (HE, original magnification 200×)
Fig. 2. Histopathology and immunohistochemistry of the spindle cell lipoma of the small intestine. A) There are spindle or stellate cells with small, uniform, fusiform, or triangulate hyperchromatic nuclei with inconspicuous nucleoli and pale, poorly defined cytoplasm within a collagenous stroma (HE, original magnification 400×). B) Entrapment of a group of ganglionic cells of submucosal (Meissner’s) plexus (HE, original magnification 400×). C) Spindle to stellate tumour cells are diffusely positive for vimentin. This stain highlights the dendritic cytoplasmic processes (original magnification 400×). D) The dendritic nature of the spindle to stellate cells was also accentuated by CD34 stain (original magnification 400×)

because they may be misdiagnosed as a benign nerve sheath tumour or even a low-grade sarcoma [10]. On the other hand, SCL on CT scan and MRI has a heterogeneous appearance with mixed fat and soft tissue density, raising a radiological concern for a mixed mesenchymal tumour or a liposarcoma [11, 12]. Thus, the correct diagnosis is made by histopathological examination and immunohistochemical study and confirmed with the finding of loss of retinoblastoma protein expression [13].

Cytogenetic analysis of SCL, pleomorphic lipoma, mammary type myofibroblastoma, and cellular angiofibroma have shown that these neoplasms share consistent cytogenetic abnormalities including deletion of the long arm of chromosome 13 (monoallelic deletion) [13].

The presence of abundant dendritic cells in our case raises the possibility that it may be a dendritic fibromyxolipoma [14, 15, 16, 17]. However, the stroma was collagenous, not myxoid, and the characteristic small plexiform vascular proliferation was not observed. Thus, the case reported herein shares cellular features with dendritic fibromyxolipoma. On the other hand, dendritic fibromyxolipoma is considered a rare variant of SCL [16].
Spindle cell lipomas are benign tumours with no propensity for local recurrence or aggressive behaviour, and conservative local excision is considered curative.

In conclusion, SCL is reported herein for the first time. The present case was clinically silent due to its small size. The histological low-fat variant is very infrequent. A particular aspect of the present case is the prominence of dendritic cells. Awareness of this histologic variant can help to avoid diagnostic difficulties. Spindle cell lipoma, although rare, should be included among the benign mesenchymal lesions of the small intestine. This report extends the range of sites in which this tumour can be found.

The authors declare no conflict of interest.

References


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