A Case of Solitary Fibrous Tumor of the Cheek

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We report a rare case with a solitary fibrous tumor of the cheek. A 73-year-old male with cheek swelling referred to our hospital. At the initial visit, a movable tumor measuring 20 × 15 mm was palpable. Therefore, the patient underwent excisional biopsy of the tumor under general anesthesia. Immunohistochemical staining showed the tumor cells to be positive for CD34, without organization of spindle cells into any discernible pattern, leading to a diagnosis of solitary fibrous tumor. The patient is now being carefully followed. There has been neither recurrence nor metastasis for 6 years, to date.

Key words: solitary fibrous tumor, cheek, CD34, pathological malignancy

INTRODUCTION

Solitary fibrous tumor (SFT), first reported by Klemperer in 1931, is a rare neoplasm, most commonly arising in the pleura [1]. SFT is now considered to be derived from mesenchymal cells, although it was previously proposed to have a vascular endothelial cell origin [2]. SFT is characterized by a patternless pattern of spindle-shaped cells and shows a high rate of positivity for CD34 on immunohistochemical staining.

SFT rarely develops in the cheek and has been reported in a few cases [3–5]. In addition, SFT may be associated with recurrence and metastasis, and some cases are found to be histopathologically malignant [6, 7].

We report a case of SFT in the cheek that was suspected to be histopathologically malignant; however, definite diagnosis was not possible in this case.

CASE REPORT

A 73-year-old man presented to a local hospital with a 1-month history of a mass in the left cheek. At the initial visit, a painless, elastic, hard, and movable mass measuring 20 × 15 mm was palpable in his left cheek. The tumor was covered with normal oral mucosa. Computed tomography (CT) showed a well-defined mass measuring 20 × 15 mm in the outer portion of the left mandible, with the same density as the muscle. Contrast-enhanced CT revealed heterogeneous enhancement inside the mass (Fig. 1). Magnetic resonance imaging (MRI) showed low signal intensity on T1-weighted image and mixed low to high signal intensity on T2-weighted image (Fig. 2) inside the mass.

The patient underwent excisional biopsy of the tumor under general anesthesia. The buccal mucosa was incised to remove the tumor along with a layer of the surrounding tissue. The removal was relatively easy, because there were no adhesions between the tumor and the surrounding tissues. The tumor was solid with internal bleeding.

Immunohistochemical staining demonstrated that the tumor cells were positive for CD34, leading to a diagnosis of SFT. However, bleeding was observed inside the tumor and some tumor cells were CD34 negative, raising suspicion of a histopathologically malignant SFT. The margin of excision was histopathologically negative; therefore, additional resection was not performed and the patient was placed under intensive follow-up. Neither recurrence nor metastasis has been observed for 6 years, to date, since the surgery.

Histopathological findings: The well-defined tumor with internal bleeding showed a patternless pattern mixed with fascicular formations and a spiral arrangement of spindle-shaped cells, as well as high cell density growth. Anisokaryosis were observed. Immunohistochemical staining showed that the tumor cells were partially positive for CD34, positive for vimentin, and 5% of the cells were positive for MIB-1. The tumor cells were negative for S-100, keratin, desmin, C-kit, and SMA (Fig. 3, 4). Based on these findings, the mass was diagnosed as an SFT.

DISCUSSION

An SFT, which develops in the soft tissues of the whole body, consists primarily of spindle-shaped cells. SFT has been difficult to diagnose and, occasionally, is confirmed by re-examining the specimen several years postoperatively [8]. There was a controversy about whether SFT was derived from mesothelial or mesenchymal tissue since the first report by Dalton et al. in 1979 [9]. However, Goodlad et al. reported eight cases in 1991, in which SFT developed in areas other than the pleura. Thus, SFTs are now considered to be derived from mesenchymal tissue because these tumors develop in areas lacking mesothelial cells [2].

Yokoi et al. reported that SFT commonly develops
Fig. 1 Computed tomography (CT) shows a well-defined mass measuring 20 × 15 mm in the external portion of the left mandible. Contrast-enhanced CT shows an internally heterogeneous mass.

Fig. 2 Magnetic resonance imaging shows a mass with mixed low to high signal intensity on T2-weighted image.

Fig. 3 The tumor is well defined (left, loupe image). The tumor shows internal bleeding (bottom right, HE staining × 40). The tumor cells have a patternless pattern mixed with fascicular formations and a spiral arrangement of spindle-shaped cells, with high cell density growth. Anisokaryosis are observed (top right, HE staining × 200).
in individuals aged between 60 and 70 years, with a moderate predominance in females, although there is no gender difference [10].

The prevalence rate is 2.8 cases per 100,000 population. The common sites include the peritoneum, retroperitoneum, pelvis, and mediastinum, in addition to the pleura.

Satomi et al. summarized clinical data in the English literature of 38 reported cases of SFT originating from the cheek [3]. Furthermore, Künzel et al. reported that the incidence of SFT originating from the cheek in the head and neck area was approximately 15% [4].

Freiser et al. reported a 12%–15% incidence of SFT in the head and neck; however, the incidence in the cheek is considered to be approximately 2% [5]. Therefore, SFT originating from the cheek was considered rare.

In general, diagnostic imaging of an SFT shows low density equivalent to that of the muscle on CT, and a contrast enhancement is often observed [11]. As the tumor grows, it shows lobation and is associated with cystic degeneration or calcification, thereby causing heterogeneous enhancement of the inside tumor. MRI often shows low signal intensity on both T1- and T2-weighted images, and contrast-enhanced imaging often shows an internal heterogeneous enhancement [11]. In the present patient, the T2-weighted image showed mixed low to high signal intensity. These imaging findings appeared to be attributable to bleeding within the tumor.

Pathologically, a patternless pattern is characteristic of SFT. Spindle-shaped cells are randomly dispersed in the densely collagenous fibrous stroma, and cell-rich areas and cell-poor areas overlap each other, with a perithelium-like vascular structure being visible.

In the area of high cell density, an MFH-like storiform pattern or a fibrosarcoma-like herringbone pattern has been reported [10]. SFT expresses CD34 at high rates since the report by Renshaw et al. in 1994 about the relationship between SFT and CD34 [12].

SFT is often positive for CD34. However, there are some soft tissue tumors that test positive for CD34; therefore, differential diagnosis is necessary. CD34-positive tumors other than SFTs include neurofibromatosis, neurilemmoma, angioleiomyoma, and hemangiopericytoma. Because hemangiopericytomas are often positive for SMA and desmin, and neurilemmomas are often positive for S-100, we diagnosed the present patient as having an SFT.

In contrast, Briselli et al. and England et al. reported that 38 (13%) of 289 patients and 82 (37%) of 223 patients, respectively, with SFTs, had clinically or pathologically malignant lesions [13, 14]. Patients with pathologically malignant SFTs are characterized with histologically increased cell density, nuclear atypia, mitosis, necrosis, ill-defined circumference, and the surrounding invasion, in contrast to benign SFTs. Van et al. and Yokoi et al. also reported that pathologically malignant SFTs were occasionally negative for CD34 [6, 7].

In our present patient, as the tumor showed internal bleeding and mitosis and was partially negative for CD34 on immunohistochemical staining, the possibility of a pathologically malignant SFT was considered. However, no recurrence, histological interstitial infiltration, and necrosis were noted. Hence, we were unable to arrive at a definitive diagnosis.

Surgery is generally the first choice of treatment for SFT [4]. The treatment for patients with a histopathologically malignant SFT may also follow this strategy. Even pathologically benign SFTs occasionally recur, such that radical surgery may greatly affect the possibility of recurrence. In our present case, the margin of excision was negative. However, considering the possibility of future recurrence or metastasis, a strict follow-up is particularly important.

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CONFLICT OF INTEREST

None of the authors have any conflict of interest to be reported.

REFERENCES


