Case 386

3. Desmoid tumor

【Progress】

He is scheduled to receive surgical mastectomy near future by oral surgeon.

【Discussion】

Desmoid is known to occur in an abdominal cavity including abdominal wall, peritoneum, and retroperitoneum, especially after surgical repair (1, 2). Desmoid can arise from intraperitoneal scars. In the process of repairing when or where fibrocytes and myo-fibrocytes work, first, fibrocytes make fibrosis for reparation. Fibrosis made by fibrocytes can be absorbed. Meanwhile, the large repair is required, second, myo-fibrocytes are followed by fibrocytes. Thick fibrosis by myo-fibrocytes cannot be absorbed, remaining permanent (2). During formation of thick fibrosis, desmoid emerges. Desmoid is considered to arise from mutation of myo-fibrocytes (2).

Desmoid also emerges sporadically and in other sites such as chest wall, shoulder, and extremities, but rare in maxillary sinus (1). The occurrence incidence of desmoid is approximately 3% of all soft tissue tumors (1). Desmoid is locally aggressive called aggressive fibromatosis, but distant metastases rarely happen. Desmoid growth accelerates with estrogen. It emerges predominantly in women rather than men under ages of 40, and over age of 40, its occurrence incidence becomes even between men and women (1, 2).

Desmoid is composed of cellular components, fibrosis and myxoid matrix. Of these, fibrosis occupies most in desmoid. The imaging of desmoid reflects on quantity of histologic components. The more fibrosis component is greater, the lower signal intensity on T2WI becomes such as ovary fibroma or uterus myoma. Further, enhancement effect becomes less because least need of vascularization on fibrosis. In our case, desmoid is visualized low signal intensity on both T1WI and T2WI probably because of the great component of fibrosis. However, cellular components and stroma matrix are greater, the signal intensity could be changed into a mix of low and iso signal intensity (3).

Desmoid can be slightly high signal intensity on Diffusion WI. ADC values are reported to be 1.493, while those are 0.873 on malignant soft tissue tumors (3). In our case ADC values were 1.006, indicative of malignancy, needing to be removed.

The treatments of desmoid are surgical resection, radiation, chemotherapy and non-steroid anti-inflammatory drugs (NSAID) (4-7). Desmoid is reported to often recur after surgical resection, indicative of being local aggressive (4). After surgical treatment, radiation and chemotherapy are reported to lessen incidence of recurrence (5, 6). When desmoid is localized and no symptomatic, the use of NSAID (meloxicam) can be possibly given since complete response for desmoid is reported(7).

In our case, desmoid tumor of maxillary sinus is very hard to obtain enough histologic specimen, surgical resection of large fields are scheduled, despite possible recurrence after surgical management.

【Summary】

We presented a sixty-one-year-old male with desmoid tumor of left maxillary sinus. The tumor was aggressive, destructing the left maxillary bone. The tumor was hypo-vascular, low signal intensity on both T1WI and T2WI. It is borne in mind that desmoid tumor arises from mutation of myo-fibroblast, inducing thick fibrosis, and it is called aggressive fibromatosis, indicative of local aggressive least with distant metastases. Although ADC values of desmoid were 1.493, those of the present desmoid were 1.006, indicative more aggressive or malignant potential, implying desmoid tumor has a wide range from malignant potential to benign character. For small desmoid tumor with less aggressive character, watchful observation using NSAID (meloxicam) is recommended.

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