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CT FINDINGS OF MALIGNANT FIBROUS HISTIOCYTOMA

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Purpose: To define the sites and CT findings of 45 MFH patients. Materials and Methods: The primary sites were : cranio-facial 16, trunk and extremity 12 and abdominal 17 (13 retroperitoneal). As seen on CT scans, the lesion is clear-margined and even when small, but, when large (42/50, >5 cm), it is uneven in consistency and apt to invade the nearby organs (75.8%). The CT findings of untreated and recurrent MFH are similar. Results: Enhanced CT scans may provide useful information such as medium to hyper-attenuation (78.8%), necrosis (60.6%) or involvement of para-nasal sinuses as expanding deformity (8/9 cases). Conclusions: CT scan is important to delineate the extent of MFH lesions of which the diagnosis is best ascertained by combining CT scan and histopathology. Enhanced CT scans can reveal much useful information.

Key words: Fibrous, Histiocytoma, Radiography, Mesenchymoma, CT scan

Malignant fibrous histiocytoma (MFH), a common malignant tumor originating from the mesencymal tissue in the adults and elderly, usually occurs in the extremities, trunk or the retroperitoneum. So far, there has been only a few reports on the image findings of sizable collections. In this report, we wish to present the CT findings of 45 MFH cases.

CLINICAL MATERIALS AND METHODS

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General Clinical Data

A retrospective review of CT findings of 45 patients with pathologically proved MFH seen between September, 1981 and December, 1994 in the Cancer Hospital, Chinese Academy of Medical Sciences was done, 26 male and 19 female. The ages ranged from 19 to 82 years with a median of 46 years. There were 27 untreated cases and 22 recurrent cases (23 episodes including four patients who had been treated for five times in our hospital). The frequency of recurrence was once to twice. The interval time between the operation of primary tumor and recurrence was 1 month to 8 years, with a median of 6 months.

The primary Sites of Tumor

In 16 patients, the tumor arose in the cranifacial area: paranasal sinus 9, infratemporal fossa 4 and one in each of nasal cavity, external auditory meatus and cranial bone. In 12 patients, the tumor originated in the soft tissue of trunk and extremities, including: neck 3, chest wall 4, abdominal wall 3 and lower extremities 2. In 17 patients, the tumor originated in the abdomen: retroperitoneum 13 (including the adrenal 3), and omentum or mesentery 4.

The Clinical Symptoms

The clinical symptoms vaied widely. Patients with cranifacial lesions: local mass 8, facial tingling 5, exophthalmus 4, head-face pain 4, impaired vision 3, nasal obstruction 2, ear plugging 2, toothache 2, nasal

discharge 2 and bloody nasal discharge 1. Symptoms of patients with lesion in the soft tissue of trunk and extremities: local mass 11 and one in each with pain in the chest wall or shoulder and hand tingling. In patients with intraabdominal lesion: mass 10, abdominal pain 5, abdominal distension 3, fever 2, and one in each with anorexia, weight loss and dysuresia.

CT Scanning Methods

CT was performed with GE 9800 HR or Somaton DRH with 5 mm collimation, 2 mm interval space in head and face or 10 mm collimation continuous in the trunk. CT Scan was obtained as 50 scanning in 45 patients, including 35 times (33 cases) on contrast material enhancement, and 29 times (28 cases) without contrast material enhancement.

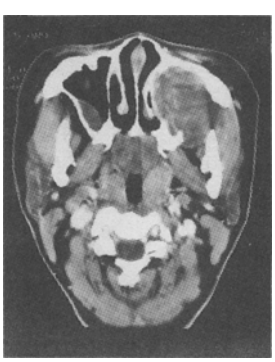
RESULTS

The Size of the Tumor and Change of Adjacent Structures on CT Scans

Of 16 patients (18 episodes) with craniofacial tumor, 15 showed a solid mass with irregular border, invading and destroying the adjacent bone wall and structures extensively. Only 1 patient with nasal lesion had a 1.5 cm mass with homogeneous density and clear margin. In 9 primary maxillary sinus lesion, the adjacent structures were obviously invaded: nasal cavity 8, palatopterygoid fossa 8, infra-temporal fossa 8, orbit 7, ethmoid sinuses 5, zygomatic bone 5 and extradural region outside the skull 3. All 4 cases originating from the infratemporal fossa invaded and destroyed the posterior wall of the maxillary sinus. Two of the four had invaded into the sinus. In three cases, the palatopterygoid fossa and pterygoid plate showed involvement. Three others also showed invasion of the nasopharynx and paraphanyngeal spaces. In 9 lesions originating from the paranasal sinuses, the sinus cavity demonstrated expansion deformity in eight. Of 12 cases with MFH in the soft tissue of the extremities and trunk, 4 originated from the chest, 3 from the neck, 3 from the pelvis and 2 from the extremities. Soft tissue mass was showed in all 12 patients with the mass ranging from 2 to 18 cm in diameter and a median diameter of 10 cm. The tumor was irregular in shape and had no definite border in 9 cases. One case had a few masses scattered. Two cases showed invasion of ribs and sternum. Two cases invaded the ilium in the pelvis. Of the 17 cases with lesion in the abdominal cavity, 13 had irregular contour, the size <10 cm 5, >=10 cm 6, >=20 cm 2, and in 6 cases the tumor could not be measured because they were too bulky. The lesion in 12 patients invaded the nearby organs: including intestine 7, kidney 2 and each of the following, liver, stomach, bladder and psoas muscle. A patient had tumor thrombus in the inferior vena cava and another had metastases in the abdominal wall.

Shape and Density of MFH on CT Scans

In 26 out of 28 cases (29 times) without contrast material enhancement, the shape of tumor was irregular and ill-defined. In 23 cases, the tumor invaded the nearby structures and organs or without clear demarcation with the latter. The consistency within the tumor was heterogeneous in 24 cases, but in 15 cases hypoattenuated necrotic area was observed inside. One case had irregular and mottled calcification. In 33 cases (35 episodes) with contrast material enhancement, the shape and border of all tumors were clear. The density of the tumor was more heterogeneous in 16 patients as compared with those having enhancement. There were apparent medium to markedly hyperattenuated enhancement in 26 cases (27 times). Necrotic areas were evident in 20 cases (21 scans). In this group, 4 cases (5 episodes) with recurrent tumor which recurred after having been operated for primary or recurrence in our hospital, the shape, density and enhancement degree in these masses were very similar to the CT findings of their respective untreated lesions (Figure 1-9).



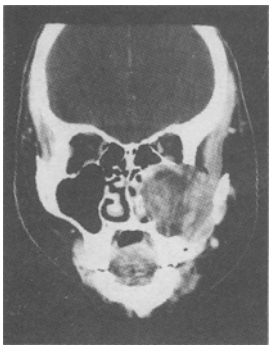
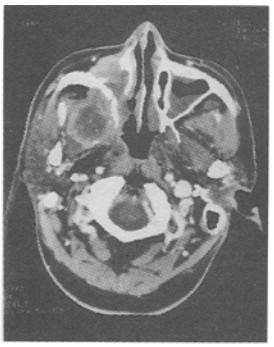


Fig 1,2. MFH of left maxillary sinus in a 26 year old female with face pain and nasal discharge for four months. The axial (1) and coronal (2) CT scan show mass in left maxillary sinus, as expanding deformity involving and destroying anterior, posterior, upper, lower, external walls invading into infratemporal fossa and facial soft tissue.



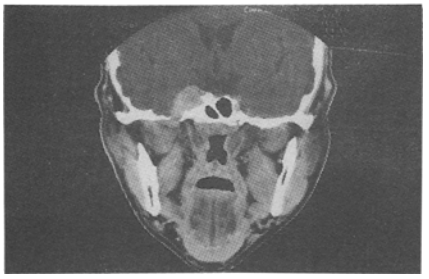


Fig 3,4. MFH of right infratemporal fossa invading into skull in a 42-year-old male with right face pain for 3.5 years. The axial(3) and coronal (4) enhanced CT scans show an irregular mass originating from infratemporal fossa which destroyed posterior wall of maxillary sinus, pterygoid plate and palatopterygoid fossa, invading into the sinus, and through the foremen oval into the cavernous sinus. The tumor showed medium enhancernent with large areas of necrosis inside.

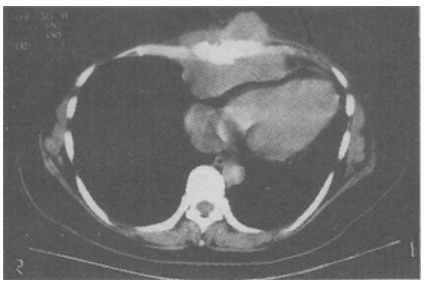


Fig 5. MFH of anterior chest wall. Image was obtained in a 46 year old women who had a history of thymoma removed and locally treated with radio-therapy eight years before in an other hospital. She was referred for a mass on the chest wall with local pain for four moths. CT scans showede an irregular mass striding across the stemum, with ill-defined margins destroging the sternum and comperessing on the pericardium.

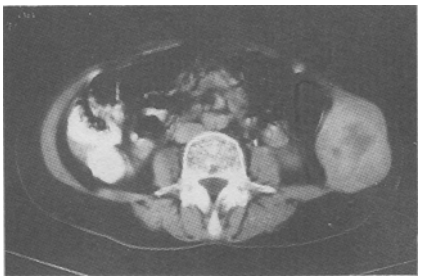


Fig 6. MFH of left lower abdominal wall in a 35 year old woman. She was referred for a huge mass on the left lower abdominal wall and buttock. Enhanced CT show a soft tissue mass in the muscle of the left lower abdominal wall with marked enhancement near the wall margin with irregular necrotic area inside.

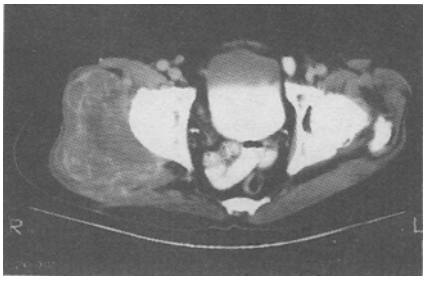


Fig 7. Local recurrent MFH of right buttock in a 79 year old male. The mass had been removed before in another hospital. Enhanced CT scans show a huge soft tissue mass in the muscle of the buttock outside of the pelvis with medium enhancement and heterogeneous density due to necrosis inside.

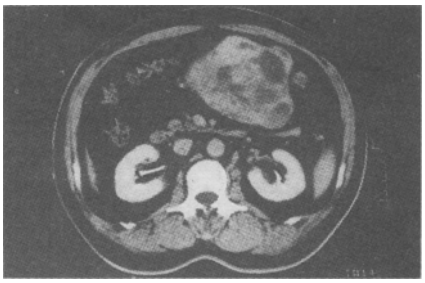


Fig 8. MFH originating from the mesentery of the transverse colon in a 55 year old male with abdominal mass for four months. Enhanced CT scans show an irregular mass in the abdominal cavity with mediam enhancement and many necrotic areas.

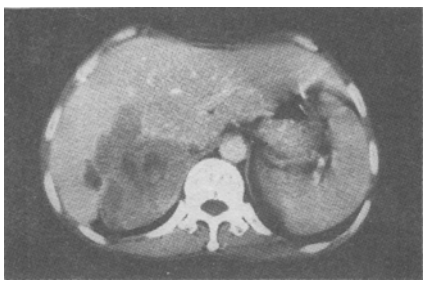


Fig 9. Right adrenal MFH invading into liver in a 58 year old male with indigestion and marked weight loss. Enhanced CT scans show a mass originating from the right adrenal gland with necrotic areas. The tumor has invaded into the liver appearing as a few irregular masses matted together.

DISCUSSION

Report on MFH were first published in 1964 by O'Brien and Stout[1] as an separate disease entity. The histopathogene origin of this tumor is believed to be amulti (pleuri)-potential cells, which differentiate towards histiocytic and fibroblastic propensities. Histitogically, the main features of this tumor are pleomorphic spindle cells, fibroblasts and histiocytes, typically arranged in sheets and fascicles (pinwheel or whirl) in storiform pattern, accompanied by giant cells and inflammatory cells. According to the character of their cytologic, histiologic appearance and quantitative in pre-dominance, it is divided into five subtypes: pleomorphic /storiform, myxoid, giant cell, inflammatory and angiomatoid. Because of the diverse appearance of this tumor, each subtype model still keeps its typical morphologic findings. MFH is a most common soft tissue sarcoma in the adults and elderly, occurring usually in the extremities, trunk and retroperitioneal space,[2-6] less commonly (3%) in the head and neck.[7] As Ros[8] stated, MFH should be first considered when seeing soft-tissue tumor in the extremities, trunk and retroperitioneal space in a person above 45 years of age. Eeles, et al. reported MFH comprising 23% of 130 cases of soft-tissue sarcoma in the head and neck area.[9] In our series, 16 cases occurred in the craniofacial area comprising 35%. This high frequency may due to the special aggregation of patients in our hospital.

On CT scans, MFH usually appears as a mass of soft-tissue density, when the tumor is small, it has well definite border and homogeneous density. When advanced, it becomes irregular in contour and heterogeneous in density with hypo-attenuated necrotic areas (55%) and calcification (7-20%) inside.[7,10,11] In our patients, all lesions appeared as soft-tissue masses. There were lower attenuation areas in 14 cases (15 episodes) and one case had calcification in 28 cases (29 episodes) on the unenhanced CT scan. Of 33 cases with enhanced CT scan, 26 (78.7%) tumors showed medium to marked enhancement. Twenty cases (60%) demonstrated lower hypoattenuated areas, which frequently appeared in the lesions in the limbs, trunk and retroperitioneal space. Of 9 patients with paranasal sinus MFH, 8 showed expanding deformity. Another characteristic of MFH is invasion of the adjacent structures or organs. In our series, 15/16 (93.8%) craniofacial MFH, simultaneously intruded into the infratemporal fossa, pterygopalatine fossa, nasal cavity, orbit or extradural region outside the skull. Of 17 patients with intraabdominal or retroperitoneal lesion, 10 (58.8%) had invasion of intestines, stomach, liver, kidney, psoas muscle or inferior evna cava at the same time. This is caused by the tumor's propensity of regional infiltration, which is striking as a special feature of its biological behavior. The pattern of spread could prompt the diagnosis of MFH. As MFH has a varied morphologic and cytological spectrum on histology, sometimes it is difficult to be differentiated from the other soft tissue sarcomas, including: 1) spindle shaped cell sarcomas, such as fibrous tissue or neurogenic tumors, 2) Sarcoma with cell shaving marked dissociation, such as pleomorphic lipossarcoma and rhabdomgo sarcoma, 3) Tumors rich in myxoid composition, such as myxoliposarcoma etc.

In this study, we have seen a few recurrent tumors of which the primary tumor had been removed at other hospitals. They had been diagnosed as having neurofibroma, neurolemmoma, angioma or leiomyosarcomas, etc. by histopathology. Therefore, a combination of electron microscopy and immunohistochemistry is necessary. The key of correct diagnosis of MFH is (1) Enough pathologic material and sections; (2) Thorough search for a criteria of tumor morphology to find the lipoblastic cells and unique changes of clump-net-like blood vessel background so as to distinguish from liposarcoma and (3) Occasional immuno-histochemical and/or electron microscopic studies are necessary. However, it is of paramount importance to closely combine the histopathologic features with the CT findings. For example, myxoid or lipoid pattern sarcomas may have fewer hyper-attenuation on CT scan when enhanced. Liposarcoma frequently gives the typical negative CT fatty value areas on the CT scans. Due to the fact that MFH being a rare tumor, we were unable to subtype our series by CT findings. Local recurrence, being most common in this tumor, is usually the chief cause of death (63%) in MFH in the head and neck region.[9] In this study, 22 cases (23 episodes) were admitted for recurrences. One patient had recurrence for 3 times which was the highest incidence in our series. The interval between the first operation and recurrence varied from one month to eight years (median six months). The CT findings of recurrent lesions were similar to those of the untreated lesion. The prognostic factors were tumor size, depth and cell grade. However, whether or not completed resection had been done first is most important.[4,6] CT scan is helpful in accurately delineating the tumor, involvement degree, extension and pattern of enhancement on the CT. These are very important for the differential diagnosis and the decision of treatment plan as well as prediction of the final outcome. CT scans without contrast material is unable to provide enough information. When the tumor has been resected, immediate CT scan should be done and periodic follow-up examinations with special reference to repeated CT scans once every year may insure a timely detection of recurrence and hence, the needed re-operation.

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