CASE REPORT

OCULAR MALIGNANT FIBROUS HISTIOCYTOMA:
A RARE CASE REPORT

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Abstract

Introduction: To date, malignant fibrous histiocytoma (MFH) is generally accepted as a type of soft tissues sarcomas. Histiocytoma originally was suspected as mesenchymal cells that cannot be differentiated. Based on previous researches, MFH is uncommon in children. This paper reports a case of ocular MFH on the management of children with chemotherapy and showed significant results.

Case Reports: A 5-year, 4-month-old boy went to Dr. Wahidin Sudirohusodo Hospital, Makassar on October 23rd 2013 with early diagnosis of suspect ocular rhabdomyosarcoma tumour. On physical examination, a lymphadenopathy (two pieces) on the left cervical glands were found with the size of each 1x1 cm, and left ocular mass with 5x5x5 cm with solid consistency. Laboratory studies revealed a white blood cell count of 8,340/mm$^3$, a haemoglobin level of 12 g/dl and platelet count 610,000/mm$^3$, renal and liver function tests were within the normal limits, diagnostic studies including radiographs of the lung showed no evidence of disease. With the diagnosis of Storiform-pleomorphic-type MFH, he was put on a combined chemotherapy with vincristine, cyclophosphamide and doxorubicin. At the fourth cycle of chemotherapy, the size of mass was decreased until no lump and the patient's eye showed normal physical appearance. No rheum on the eye or inflammation after the chemotherapy.

Conclusion: The diagnosis of ocular MFH in a 5-year, 4-month-old boy was established by history taking, physical examination and histopathological examination. The treatment was chemotherapy and the prognosis depends on the site and location of the tumour.

Keywords: Ocular Malignant Fibrous Histiocytoma, MFH

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Introduction

A case of ocular malignant fibrous histiocytoma (MFH) in a 5-year, 4-month-old boy was reported. Diagnosis based on anamnesis obtained lump on the left eye which had been observed since 6 months before entering the hospital. On physical
examination, it was found that a mass present in the left eye measuring 5x5x5 cm and histopathological examination showed malignant fibrous histiocytoma. The therapy was a combination of chemotherapy consisting of vincristine, cyclophosphamide and doxorubicin. The result showed a significant improvement. This case-report summary is a novel treatment for ocular MFH in our department.

MFH is a soft tissue sarcomas derived from primitive mesenchymal cells into the cells with characteristics manifest fibroblastic and histiocytic. MFH is classified into five subtypes: Storiform-pleomorphic, myxoid, Giant-Cell, inflammatory and Angiomatoid. Each subtype is distinguished from the arrangement of forms/types of cells [1,3]. MFH is very rare in children. It is more experienced by adults aged 50 to 70 years. From a study of 23 cases of MFH in the eye that has been discovered, with age ranged from 3 to 85 years, only two cases occurred in children (3 and 9 years old, respectively) [3]. Some literatures reported that MFH malignancies in children are relatively lower than adults and most commonly found among Caucasians (91%) than African/Asians (2-7%) [4].

MFH may occur due to radiation exposure, shrapnel injuries, fixation of the metallic equipment, prostheses joints (total joint prostheses) and even foreign substances getting into the body [5]. A clinical manifestation of MFH is generally in the form of an asymptomatic mass. Symptoms occur is mainly due to mechanical compression, traction or entrapment of the nerve fibers or muscle fibers. Clinical symptoms of MFH in the eye are quite diverse, ranging from decreased visual acuity, proptosis, diplopia, limitation of extra ocular muscle movement, swelling of the eyelids and conjunctiva [6]. MFH diagnosis can be confirmed by biopsy, magnetic resonance imaging (MRI) or computed tomography (CT) scan. More accurate results can be obtained from immunohistochemistry. Procedures in dealing with MFH can be surgical excision, surgical resection, chemotherapy, radiation and adjuvant radiotherapy [5]. Seventy-five percent of patients with MFH experienced local recurrence with approximately 30% had metastatic. MFH with 5-years survival rate can be increased with tumour resection and radical extensively, including the area around the tumour. Radiotherapy is also done in adult case. However, a controlled study of radiotherapy in children has not been found [3]. Chemotherapy becomes a more preferred alternative, especially when the body response well to chemotherapy, namely the provision combination of vincristine, cyclophosphamide and Adriamycin [6].

Case presentation

A 5-year, 4-month-old boy, went to Dr. Wahidin Sudirohusodo Hospital, Makassar, on October 23rd, 2013, with early diagnosis of suspect ocular rhabdomyosarcoma tumour. Lump on the patient's left eye had been observed since 6 months before entering the hospital. Lump originated from the lower eyelid with slowly growing and urged the eyeballs out. The patient got a febrile, but with no seizures, no cough, no mucus and no shortness of breath. He also had no nausea or vomiting. Patient experienced decreased of appetite. He felt weak, but not agitated or palpitations. The patient’s defecation was normal and yellow, while his urinating was smooth and yellow. On physical examination, a lymphadenopathy (two lumps) on the left cervical glands were found with the size of each 1x1 cm and left ocular mass with 5x5x5 cm with solid consistency. No other
remarkable findings on the physical examination were detected. Laboratory studies revealed the white blood cell count of 8,340/mm$^3$, the haemoglobin level of 12 g/dl, platelet count of 610,000/mm$^3$, renal and liver function tests were within the normal limits and diagnostic studies including radiographs of the lung showed no evidence of disease. Head CT-scan without contrast revealed an extra cranial mass on left ocular with suspect rhabdomyosarcoma. Since the mass was unresectable, a tru-cut biopsy was done for diagnosis. Bone marrow aspiration revealed no infiltration. The histopathological examination showed the presence of MFH.

With the diagnosis of Storiform-pleomorphic-type MFH, he was put on a combined chemotherapy with vincristine, cyclophosphamide and doxorubicin. At the fourth cycle of chemotherapy, he showed significant result. The size of mass was decreased until no lump and the patient’s eye showed normal physical appearance. No rheum on the eye, also no inflammation after the chemotherapy.

**Figure 1. The condition of patient in pre (left) and post (right) after fourth cycle of chemotherapy**

**Discussion**

Malignant fibrous histiocytoma (MFH) was first discovered and described by O'Brien and Stout in 1960 and Kempson and Kynakos in 1972. To date, MFH is generally accepted as a type of soft tissues sarcomas. Histiocytoma originally was suspected as mesenchymal cells that cannot be differentiated (undifferentiated) [7, 3]. Based on previous researches, MFH is uncommon in children. The incidence was found mostly in adults aged 50-70 years, in the area of extremities (68%) and retroperitoneal (16%), and very rarely in the head and neck (9%) [3]. Prevalence of MFH incidence in children ranged 2-7% cases of child sarcoma [8-10]. Comparison of incidence between boys to girls is 2:1 [9-10]. MFH in ocular neoplasm is composed of fibrotic and histiocytic elements [6]. Histiocytic and fibroblastic pattern contained in paediatric patient’s tumour cells, resembling the tissue neoplasm in adults [10].

MFH ocular diagnosis in this case based on history, physical examination and investigations. In anamnesis obtained a 5-year-old boy came with a lump in the left eye observed since 6 months before entering the hospital. On physical examination, a mass was found in the left eye measuring 5x5x5 cm. The investigations based on the head CT-scan, we found that there were heterogeneous mass of 4.2x5.3 cm in size in
extra cranial, well defined, destruction surrounding bone and extended to the bulbous oculi anterior urgent, out of the orbital cavity, sinuses maxillaries left and to the lateral left zygomatic area with extra cranial mass impression left orbital rhabdomyosarcoma suspected. However, after a biopsy was done, we concluded that it was a malignant fibrous histiocytoma. When viewed microscopically, the tumor cells were round oval shape, vesicular core with quite prominent nucleoli, mitotic pretty much and in the arrangement of storiform. Therefore base on the mentioned findings, MFH in this case was classified as pleomorphic subtype-storiform. This subtype is characterized by a mixture composed of spindle cells with round or polygonal shape in the storiform pattern. There can also be multinucleated giant cells, but in general it can be characterized by an abundance accompanied nuclear pleomorism atypical mitosis. In addition, this subtype has the highest prevalence of cases of MFH, which account for about 60-70% [1, 10] (Level IV Recommendation E).

Figure 2. Tumour composed of arranged storiform cell with prominent nucleoli, mitotic quite a lot, a little necrosis (hematoxylin and eosin-HE, X 40)

Figure 3. Head CT-scan revealed an extracranial mass on left ocular
Primary therapy for MFH may include surgery, radiation therapy or chemotherapy. Surgery, particularly extensive surgical excision (surgical excision wide) [9], although it can be done, it has a high local recurrence radiation therapy to the size of 4000-6000 rad and it is able to improve survival in patients with soft tissue sarcomas and substantially reduce the level of recurrence in adult patients with MFH [10]. Radiotherapy has been used in adults cases, but there are no reports in paediatric cases yet [6]. Similarly, although there has been no research on the effectiveness of the MFH, but still recommended to metastatic disease or tumor that cannot be resected again [9]. The combination of vincristine, cyclophoshamide and adriamycin was advised to treat other soft tissue sarcomas. Some researchers also suggested to use ifosfamide rather than cyclophoshamide [9, 6] (Level V Recommendation E) and it was applied in the case of our patient (M).

A research that was conducted by Çağlar et al. also suggested that the survival rate for 5 years is influenced by the size of the tumour, i.e. <5 cm by 82%, 5-10 cm by 63% and >10cm by 51% (Level IV Recommendations E). In addition, the level of prognosis in children with MFH is better than adult [9]. Therefore, follow-up till more than 10 years is needed due to some local recurrence or distant metastases may occur in the future [6].

**Conclusion**

The diagnosis of ocular malignant fibrous histiocytoma in a 5-year, 4-month-old boy was established by history taking, physical and histopathological examinations. Chemotherapy is the recommended treatment and the prognosis is depending on the site and location of the tumour.

**References**


