Post operative radiation and chemotherapy for R1 resected Solitary fibrous tumor at nasal cavity and paranasal sinus: A case report

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บทคัดย่อ

เนื้องอกชนิด solitary fibrous tumor ในบริเวณศีรษะและลำคอพบได้ค่อนข้างยากโดยเฉพาะอย่างยิ่งในตำแหน่งโพรงจมูกและโพรงไซนัส การรักษาหลักสำหรับเนื้องอกชนิดนี้ก็คือการผ่าตัด และเพื่อที่จะให้ได้ผลการรักษาที่ดีนั้น จำเป็นที่จะต้องได้การผ่าตัดที่เพียงพอ สำหรับการรักษาเสริมด้วยการฉายรังสี ในกรณีที่ผ่าตัดออกไม่หมด ยังเป็นที่ถกเถียงกันอยู่ รายงานผู้ป่วยเรื่องนี้เป็นการรายงานกรณีผู้ป่วยชายอายุ 49 ปี ที่มี solitary fibrous tumor ที่โพรงไซนัสด้านซ้าย โดยได้รับการรักษาด้วยการผ่าตัดและมีผลชิ้นเนื้อระบุว่าผ่าตัดออกไม่หมด ดังนั้นผู้ป่วยรายนี้จึงได้รับการฉายรังสีหลังผ่าตัด 6600 เซนติเกรย์ และเคมีบำบัดตามหลังด้วย Doxorubicin, Ifosfamide และ mesna จำนวน 6 รอบ หลังจากฉายรังสีครบ 3 ปี ผู้ป่วยยังคงปราศจากโรคกลับเป็นซ้ำ และโรคแพร่กระจาย

Abstract

Solitary fibrous tumor of head and neck region is quite rare especially in nasal cavity and paranasal sinus location. The main treatment modality for this tumor is surgery. To achieve good clinical outcomes, adequate resection is needed. The role of adjuvant radiation, in case of inadequate resection, is still debated. We present a case of a 49-year old male who had solitary fibrous tumor at left maxillary sinus. He underwent total maxillectomy and his pathological reports showed positive margin. He then received post-operative radiation at a dose of 6600 cGy and 6 cycles of chemotherapy consisted of doxorubicin, ifosfamide and mesna. Three year after complete radiation, he remained without evidence of local recurrence and distant metastasis.

Introduction

Solitary fibrous tumor is an uncommon spindle cell tumor which originates from mesenchymal cell. It was firstly described by Klempere and Rabin in 1931. The most common site of this tumor is pleura. However, it can occur in extrapleural regions such as liver, abdomen, extremities, urogenital tract and head and neck. For head and neck, solitary fibrous tumor was reported less than 0.1 % of all upper respiratory tracts tumor. Moreover, it was rarely found in nasal cavity and paranasal sinuses[1,2,3]

It usually presents with local compressive symptoms which is difficult to distinguish from other soft tissue tumor[1]. The radiographic finding of this tumor are nonspecific. Therefore, the diagnosis may depend on histopathologic, cytologic and immunophenotypic features. Most cases show proliferation of spindle cell and ovoid cell in vascular and collagenizedstroma. All cases are positive for vimentin, CD-34, BCL -2 and CD-99[4,5].

For treatment, adequate surgery with negative margin leads to favorable outcome. In patient who cannot achieve clear margin, adjuvant radiation is still debated[6]. So, we reported extremely rare case of solitary fibrous tumor and has a good result from multimodality treatment.

Case Report

A 49-year-old Thai male presented with mass at left cheek. She underwent computerized tomography of head and neck region. The study found large rim-enhanced mass (size 5.5 x 6.7 x 5 cm) in left maxillary sinus with expansion and bony destruction at lateral wall as well as left zygoma without lymphadenopathy. His computerized tomography of the chest was normal. After that, the patient underwent biopsy and the pathological report showed spindle cell lesion, suggestive of solitary fibrous tumor (positive for CD-34, BCL-2, CD-99, but negative for S -100 and desmin) moderate to marked pleomorphismhypercellular frequent mitosis 4/10. Therefore, he was treated with total maxillectomy and the pathological finding corfirmed malignant solitary fibrous tumor 5 x 4.5 x 3.8 cm, no lymphvascular invasion, positive medial, lateral and superior maxillary sinus margin. After surgery, he received post operative three dimensional radiotherapy at dose of 6000 cGy to tumor bed and 6600 cGy to area of positive margin. He was then treated with 6 cycles of chemotherapy consisted of doxorubicin, ifosfamide and mesna. After complete radiotherapy for 3 year, he remained without evidence of locoregional recurrence or distant metastasis. He had minimal trismus, dry eyes and dry nose.

Discussion

As we know, solitary fibrous tumor is a tumor which originated from mesenchymal cell and we can find it in any site that have serosa. The most common site of solitary fibrous tumor is pleura[1,2]

The retrospective study of Bowe et al.[1] examined about solitary fibrous tumor in head and neck and showed that all of solitary fibrous tumor at nasal cavity presents with nasal congestion. The other symptoms are epistaxis, rhinorrhea, headache and facial pain.1 Furthermore, the comparative study of Gold et al.[7] revealed that patients with extrathoracic solitary fibrous tumor had less symptoms at presentation than patients with thoracic solitary fibrous tumor. 7 To diagnosis solitary fibrous tumor, we can perform endoscope and Computerized Tomography scan (CT scan) or Magnetic Resonance Imaging (MRI scan). 2 Ganly et al. found that 80% of CT scan findings of solitary fibrous tumor at head and neck had homogenous enhancement, which were different from CT scan findings of solitary fibrous tumor at non-head and neck area according to study of Wignall et al[1,8,9]. In addition, MRI scan of solitary fibrous tumor demonstrated isosignal intensity to muscle and brain in T1 and T2-weighted images.

For pathological diagnosis, there is difficulty due to its various characteristics like other soft tissue tumors. It is usually found proliferation of spindle cell, ovoid cell in vascular and collagenisedstroma. 4,5 And mostly solitary fibrous tumors are positive vimentin, CD-34, BCL-2 and CD 99. 2,4,5,6

The main treatment modality to treat solitary fibrous tumor is surgery. And the most importance factor which impact to clinical outcomes is

resectability; the. more adequate surgery, the better clinical outcomes. A retrospective study of Bowe et al.[1] revealed that all patients underwent surgery and 10 of 13 patients had pathologically clear margins. Only one patient treated with post operative radiation. However, there was no local recurrence among these patients. 1 Gold et al. examined about correlation of clinicopathologic and solitary fibrous tumor. They found that factors affected local recurrence significantly in resectable solitary fibrous tumor are recurrence on presentation, extrathoracic location, macro/microscopic positive margin, more than 4/10 PF of mitoses, necrosis and presence of malignant components. They also recommended that adjuvant radiation should be considered in case of tumor size more than 5 cm or inadequate surgical margin.7 For this case, tumor size was 5 cm and surgical margins were positive medial, lateral and superior. In addition, there were nuclear pleomorphism, area of increased cellularity and mitotic index 4/10 HPF in this case , which were indicated malignant features and affect tumor recurrence. So we considered giving post- operative radiation to the tumor bed.

Furthermore, combined surgery and radiation can contribute excellent local control in solitary fibrous tumor. Bishop et al. investigated about survival outcomes and local control in solitary fibrous tumor patients who treated with surgery and radiation. There were solitary fibrous tumor at head and neck area about 32 % including in this study. Forty-five percent of patients were treated with post -operative radiation (median dose of 58 Gy) .

The study showed that 5-year local control, overall survival and distant metastasis free survival were 100%, 95% and 92%, respectively. They suggested to treat patients with 50 Gy of preoperative radiation or 60 Gy of post-operative radiation using conventional fractionation. In case of positive margin, they did not recommend dose escalation beyond 60 Gy[10].

Regarding the role of chemotherapy, there is a retrospective study of Stacchiotti et al analyzed about response of anthracycline-based chemotherapy with or without ifosfamidein solitary fibrous tumor. The study reported that median progression free survival was 4 months, which median progression free survival of dedifferentiated subtypes was much more than malignant subtypes (5 months and 3.5 months respectively). They also implied that dedifferentiated solitary fibrous tumor has better response to chemotherapy in their series[11]

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