Computed Tomography of Lethal Midline Granuloma

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— Abstract —

In order to clarify the CT findings of lethal midline granuloma (LMG) diagnosed clinically or histopathologically, the authors retrospectively analyzed 12 patients who were seen at Kyungpook National University Hospital from February 1985 to August 1989.

CT showed nasal mucosal thickening and/or soft tissue mass (9 cases), spreading of the lesions along the facial subcutaneous fat plane (8 cases), invasion into the paranasal sinuses (5 cases), bone destruction (5 cases), nasopharyngeal mass lesion (2 cases), and extension of the lesion into the infratemporal fossa (1 case).

In spite of the fact that CT does not make definitive diagnosis of LMG, it permits evaluation of the extent of the lesion, detection of the combined lesion, differential diagnosis, and close monitoring of its evolution under treatment.

Index Words: Nose, Lethal Midline Granuloma 261.622
Paranasal sinuses, Computed Tomography 23.1211
Nose, Computed Tomography 261.1211

Introduction

Since the first report of McBride (1897)(1), the term lethal midline granuloma (LMG) and its various synonyms—progressive lethal granulomatous ulceration (Stewart, 1933), lethal granulomatous ulceration of midline facial tissue (Williams, 1949), idiopathic lethal granulomatous ulceration (Woodburn & Harris, 1951), nonhealing granuloma (Waltone, 1959), polymorphic reticulosis (Eichel et al., 1966), midline malignant reticulosis (Kassel et al., 1969), midline granuloma (Dellon, 1977), and lymphomatoid granulomatosis (DeRemee et al., 1978)—have been used to describe a rare lesion of a nonhealing granuloma that ultimately causes necrosis and destruction of the nose and midface. According to WHO (1978), LMG is defined as a progressively destructive nontuberculoid granulomatous lesion localized in the nasal cavity, paranasal sinuses, or palate and infiltrating the soft and hard tissue.

Recent research on the condition historically known as lethal midline granuloma concludes that it is a form of T-cell lymphoma.(2)

The prominent histological features of LMG are inflammation, necrosis, and thrombosis with infiltration of the atypical histiocytes into the perivascular connective tissue.

The purpose of this retrospective study was to clarify the CT findings of LMG.

Materials and Methods

Our study included 12 patients with LMG who were seen at Kyungpook National University Hospital from February 1985 to August 1989. They were 9 men and 3 women with an age range of 17 to 71 years (mean: 40 yrs).

Four cases were pathologically proved and 8 cases had nonspecific pathologic findings, but their clinical findings and courses were identical to LMG.

All patients were examined with a CT/T 8800
Results

The most common clinical features of LMG was nasal stuffiness. Other symptoms and signs included facial swelling and pain, mucocutaneous ulceration, nasal discharge, and foul odor (Table 1). The primary lesion sites were the nasal cavity (8 cases), nasopharynx (2 cases), palate (1 cases), and external nose (1 case).

The main histological findings were acute or chronic inflammation (12 cases) and focal or massive necrosis (7 cases). Atypical histiocyte infiltrations

Table 1. Clinical Features

<table>
<thead>
<tr>
<th>Symptoms/Signs</th>
<th>No. of cases</th>
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<tr>
<td>Nasal stuffiness</td>
<td>8</td>
</tr>
<tr>
<td>Facial swelling and pain</td>
<td>5</td>
</tr>
<tr>
<td>Mucocutaneous ulceration</td>
<td>4</td>
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<tr>
<td>Nasal discharge</td>
<td>3</td>
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<tr>
<td>Swallowing difficulty</td>
<td>2</td>
</tr>
<tr>
<td>Sore throat</td>
<td>2</td>
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<tr>
<td>Foul odor</td>
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scanner (GE, Milwaukee, WI, U.S.A.) using contiguous 5 or 10-mm-thick sections from the frontal sinus to the hyoid bone after 50cc bolus and 100cc dripping of contrast (Rayvist®300) administration.

Axial or axial with coronal images were obtained.

Fig. 1. a, b. A soft tissue mass lesion is seen in posterior portion of the left nasal cavity. Nasal septum is perforated. The mucosa of the anterior nasal septum is mildly thickened, and the lesion spreads along the left facial fat plane. Soft tissue of slightly lower density in left maxillary sinus suggests inflammation secondary to obstruction of maxillary sinus ostium.

Fig. 2. Section through the midantrum shows soft tissue mass filling both sides of the nasal cavity. Bone destructions are present involving both medial walls of the maxillary antra.

Fig. 3. a, b. There is a soft tissue mass occupying nasal cavity with mucosal thickening. Irregular bone destructions are present in the medial wall of the right maxillary antrum. Chronic sinusitis is present in the right maxillary antrum.

Fig. 4. A soft tissue mass lesion is involving the anterior portion of the right nasal cavity and nare.
were seen in 4 cases, but fibroid necrosis, granuloma, and giant cells were not observed.

Prominent CT findings included nasal mucosal thickening and/or a soft tissue mass (9 cases) (Fig. 1, 2, 3, 4). Spreading of the lesions along the facial subcutaneous fat plane was noted frequently (8 cases) (Fig. 1, 5). Invasion into the paranasal sinuses was observed in 5 cases; the maxillary sinus (5/5), ethmoid sinus (5/5), and sphenoid sinus (4/5) (Fig. 2), but frontal sinus invasion was not seen.

Bone destruction (5 cases) and nasopharyngeal mass lesion (2 cases) were also observed (Fig. 2, 3, 4, 5). Extension of the lesion into the infratemporal fossa was seen in 1 case (Table 2).

Table 2. CT Findings

<table>
<thead>
<tr>
<th>CT findings</th>
<th>No. of cases</th>
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<tr>
<td>Nasal mucosal thickening and/or soft tissue mass</td>
<td>9</td>
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<tr>
<td>Spreading of the lesions along the facial subcutaneous fat plane</td>
<td>8</td>
</tr>
<tr>
<td>Invasion into paranasal sinuses</td>
<td>5</td>
</tr>
<tr>
<td>Bone destruction</td>
<td>5</td>
</tr>
<tr>
<td>Mass in nasopharynx</td>
<td>2</td>
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</table>

Discussion

The term "lethal midline granuloma" does not properly reflect the current observation that several different diseases can produce noninfectious midfacial destruction. In many cases, this is neither lethal nor midline, and granuloma may be absent. Recently, the disorders producing erosion of the upper aerodigestive passage are more accurately classified by clinical and histological criteria as idiopathic midline destructive disease, Wegener's granulomatosis, polymorphic reticulosis, and nonHodgkin's lymphoma.(3)

The etiology of LMG has been controversial, and the proposed pathogeneses have been immunologic disorder(4, 5), neoplastic process(6), or close relation with lymphoma(7). But recent research concludes that it is a form of T-celllymphoma(2), and the application of immunological studies with monoclonal antibody panels is now a standard procedure and should help in avoiding misdiagnosis. Kataura et al.(8) studied 3 patients with lethal midline granuloma using monoclonal antibodies against T-cell subsets, clearly demonstrating that the tumor cells in each case were probably T-cell, not from histiocytes, reticulum cells, or B-cells.

The prominent histological features of LMG are inflammation, necrosis, and thrombosis, all 3 of which suggest a similarity to Wegener’s granulomatosis, but differentiate the lesion from lymphosarcoma(9). True vasculitis is a typical finding in Wegener's granulomatosis but does not appear in the LMG, although polymorphic cells may infiltrate the perivascular connective tissue mimicking vasculitis. Histologically, the lesions of LMG are separable from Wegener's granulomatosis by the absence of necrotizing epithelioid granulomas, a tendency toward a necrotizing angioinfiltrative growth pattern, and abundant atypical lymphoproliferative cellular components(10). In LMG cases, only ulceration of the upper respiratory tract occurs, whereas the lungs and kidneys are never affected. This specific organ involvement is also a significant point in differential diagnosis from Wegener's granulomatosis.

A recent study on the CT findings of LMG show-
ed a more or less complete filling of the air cavities of the face by an abnormal soft tissue mass reaching the nasal cavity and the nasopharynx, the ethmoidal cells, the sphenoidal sinus, the maxillary sinus, and the frontal sinus(11). These findings are identical to the results of our study, except for the frontal sinus involvement. CT permits the precise evaluation of the extent of the lesions. It is known that the lesions can extend to the intracranium, to the infratemporal fossa, and to the orbit and can infiltrate the subcutaneous tissue of the face(11). In this study, cranial or orbital invasion was not observed, but the spreading of the lesion along the facial subcutaneous fat plane was frequently seen. Paranasal sinus invasion was not infrequently seen, but it is difficult to differentiate it from chronic sinusitis caused by obstructive nasal lesions. Presence of major bone destruction with no definite osteosclerotic bone thickening is helpful for differential diagnosis.

The CT permits no definite differentiation of LMG from other granulomatoses or from nasal or paranasal sinus tumor lesions. Nevertheless a few differential points may be mentioned: There is no calcification within the granulomatous tissue, osteosclerotic reaction at the bone structure level, or cervical locoregional lymph nodes(11). In our study, calcification or lymph node enlargement was not observed. The nasal or paranasal sinus carcinomas tended to be bulky and expandable with nonspecific focal bone destruction(12), but these findings were rare in our series. CT is also useful in detection of combined lesions, distant metastases, or close monitoring of the LMG under treatment, which is essentially radiotherapy.

The prognosis of LMG is poor(5,13). However, there are many recent reports about its good response to radiotherapy(14,15). In our 12 patients examined between February 1985 and August 1989, 6 patients showed complete remission on follow-up studies.

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치사성 정중부 육아종의 전산화 단층촬영

이호석, 김태호, 서경진, 김태현, 김용주, 강덕식

치사성 정중부 육아종은 비장, 부비강, 비인강을 포함한 상기도 및 안면에 국소적 및 전기성 질환이며, 파괴적인 염증성 병변이 나타나는 질환으로서 예후가 아주 불량하며 1897년 McBride가 처음 기술한 이래 비록 발생빈도는 드뢰하나 수많은 학자들에 의해 여러가지 병명이 소개되고 있다. 병리소견 등에 대해 많은 연구가 진행되어 왔다.

병리 조직학적 소견이 비특이적인 경우가 많으므로 생검시에는 심부 조직 및 궤양변연부 조직을 얻어야하며 반복적 생검이 확진에 필수적이다.

저자들은 1985년 2월부터 1989년 8월까지 경북대학교병원을 방문하여 치사성 정중부 육아종으로 진단받은 12명의 환자를 대상으로 전산화 단층촬영 소견을 역행적으로 분석하여 그 특징에 대해 알아보고자 하였다.

전산화 단층촬영 소견으로는 비강점막 비후와 비강내 연부조직 종괴가 가장 흔한 소견이었고 (9례) 안면부 피하 지방층을 따라 염증성 병변이 파급되는 양상이 8례에서 관찰되었다. 부비강으로의 병변의 파급과 비특이적인 골괴화 양상이 각각 5례에서 관찰되었으며 비인강 종양과 비슷한 양상을 보인 예도 2례가 있었다. 그러나 임파선 종괴나 병변 석화화 양상은 관찰되지 않았다.