Salivary Gland Swelling in Wegener's Granulomatosis: A Rare Cause of a Frequent Symptom

VOLKER GASSLING, JÖRG WILTFANG, JOCHEN HAMPE, JAN HINRICH BRÄSEN, MARCUS BOTH and FRANK MOOSIG

J Rheumatol 2010;37;2633-2635
http://www.jrheum.org/content/37/12/2633

1. Sign up for TOCs and other alerts
http://www.jrheum.org/alerts

2. Information on Subscriptions
http://jrheum.com/faq

3. Information on permissions/orders of reprints
http://jrheum.com/reprints_permissions

The Journal of Rheumatology is a monthly international serial edited by Earl D. Silverman featuring research articles on clinical subjects from scientists working in rheumatology and related fields.
Salivary Gland Swelling in Wegener’s Granulomatosis: A Rare Cause of a Frequent Symptom

To the Editor:

Wegener’s granulomatosis (WG) is a multisystemic disease with a complex genetic background. The clinical presentation is characterized by necrotizing granulomatous inflammation of the upper and lower respiratory tract, glomerulonephritis, and small-vessel vasculitis. We describe an unusual primary manifestation of WG with salivary gland swelling.

A 68-year-old man who was admitted to the hospital presented with progressive swellings of his right cheek and bilateral submandibular space over a period of several weeks. Extraoral examination revealed a bilateral enlargement of both submandibular glands and the right parotid gland, which were solid, plain, and painless to pressure (Figure 1.1). The oral cavity showed no pathologies of the mucosa, and no putrid secretion from the Stenon’s duct was determined. His history was notable for eye complications with dryness and reddening, ear complaints with bilateral hearing impairment, severe headache, night sweats, muscular weakness, and weight loss of about 8 kg during the 6 weeks before presentation. Blood tests revealed increased values for erythrocyte sedimentation rate (110/115 mm), C-reactive protein (203.7 mg/l), and leukocytes (13.18/nl). On magnetic resonance imaging (MRI), the right parotid gland and the submandibular glands were enlarged, demonstrating low signal intensity in T1-weighted and T2-weighted imaging (Figure 1.2 a and b, and Figure 1.3).

Figure 1. 1.1: Note the massive bilateral enlargement of submandibular space and the right cheek. 1.2: Axial noncontrast enhanced T1-weighted (a) and T2-weighted, fat-suppressed sequences (b) show a markedly enlarged right parotid gland with hypointense signal. There is homogeneous enhancement in T1-weighted fat-suppressed imaging after intravenous contrast media application (c). 1.3: In contrast to T1-weighted imaging (a), T2-weighted fat-suppressed (b) and contrast enhanced T1-weighted fat-suppressed (c) sequences reveal inhomogeneous signals of the enlarged submandibular glands, leading to suspicion of presence of small necrotic parts.
a and b). After intravenous application of contrast media, marked enhance-
ment was found in the affected sites, with inhomogeneous signals in the
submandibular glands (Figure 1.2c and Figure 1.3c). There was no evi-
dence of abscess formation or malignancy in which small areas of necrosis
could be found. The patient left the clinic without a firmly established di-
agnosis and refused further diagnostic measures. He underwent medical
examinations at regular intervals by his general practitioner. A presumptive
diagnosis of Heerfordt syndrome was made (an acute syndromal presenta-
tion of sarcoidosis, presenting with fever, uveitis, and swelling of the
parotid and/or other salivary/lacrimal glands) and longterm treatment with
a reducing regime of prednisolone was started (beginning with 60 mg
daily). The swelling of the salivary glands disappeared after 3 months, but
general symptoms persisted. Meanwhile, a further laboratory analysis
revealed elevated cytoplasmic antineutrophil cytoplasmic antibodies
cANCA; 1/276) and proteinase-3-ANCA (196.0 U/ml), results that raised the
suspicion of WG. However, the patient again refused further medical ex-
aminations to confirm the diagnosis. A definite diagnosis of WG was
confirmed at autopsy by histological evaluation of renal specimens follow-
ing unexpected cardiac death about 1 year later. The autopsy showed pauci-
immune extracapillary proliferative glomerulonephritis (Figure 2).

Salivary gland swelling is a frequent symptom reflecting different pathologies. If the enlargement is not confined to 1 salivary gland, a sys-
temic cause must be assumed. In spite of that, even other diseases usually
causing unilateral salivary gland swelling may occasionally induce bilater-
al swelling. So it is appropriate to keep all these diseases in mind when
establishing an accurate diagnosis (Table 1). We observed an unusual pri-
mary manifestation of WG with bilateral enlargement of both sub-
mandibular glands and the right parotid gland. Accurate diagnosis of WG
is based on clinical, histopathological, and blood chemical investigations.
Additionally, classification according to the criteria of the American
College of Rheumatology (ACR)2 seems apt. However, there are cases in
which the ACR criteria are not fulfilled. In particular, so-called localized
WG, possibly initially present in our patient, is not considered in the ACR
criteria. According to the European Vasculitis Study Group, “localized dis-
ease” is defined as WG without vasculitis outside the ear, nose, and throat
tract and the lung, without threatened vital organ function, and without
constitutional symptoms3.

In these cases, an early cANCA test may aid diagnosis and provide
essential information for early therapy and thus prevent disease progres-
son. A disadvantage is that the sensitivity depends largely on the extent of

Table 1. Diseases accompanied by salivary gland enlargement.

<table>
<thead>
<tr>
<th>1. Infection</th>
<th>Bacterial</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Acute purulent</td>
</tr>
<tr>
<td></td>
<td>Chronic recurrent</td>
</tr>
<tr>
<td></td>
<td>Specific bacterial</td>
</tr>
<tr>
<td></td>
<td>Tuberculosis</td>
</tr>
<tr>
<td></td>
<td>Viral</td>
</tr>
<tr>
<td></td>
<td>Epidemic parotitis (rubulavirus)</td>
</tr>
<tr>
<td></td>
<td>Cytomegalovirus (cytomegalovirus)</td>
</tr>
<tr>
<td></td>
<td>Other (Kaposi sarcoma herpesvirus, Epstein-Barr virus, etc.)</td>
</tr>
<tr>
<td>2. Obstructive electrolyte sialadenitis (+/- sialolithiasis)</td>
<td></td>
</tr>
<tr>
<td>3. Sialadenosis</td>
<td>Endocrine disturbance (e.g., diabetes mellitus)</td>
</tr>
<tr>
<td></td>
<td>Psychogenic disturbance (e.g., bulimia)</td>
</tr>
<tr>
<td></td>
<td>Dystrophic disturbance (e.g., cachexia)</td>
</tr>
<tr>
<td>4. Autoimmune disease</td>
<td>Sjögren’s syndrome</td>
</tr>
<tr>
<td></td>
<td>Autoimmune pancreatitis</td>
</tr>
<tr>
<td>5. IgG4 sclerosing disease</td>
<td></td>
</tr>
<tr>
<td>6. Granulomatous disease</td>
<td>Sarcoaidosis</td>
</tr>
<tr>
<td></td>
<td>Wegener’s granulomatosis</td>
</tr>
<tr>
<td>7. Neoplastic disease</td>
<td>Benign</td>
</tr>
<tr>
<td></td>
<td>Malignancies</td>
</tr>
<tr>
<td>8. Iatrogenic</td>
<td>Radiation</td>
</tr>
<tr>
<td></td>
<td>Radio-iodine</td>
</tr>
</tbody>
</table>

Figure 2. Kidney autopsy section showing cellular crescent formation in
the right part of the glomerulus (arrow). In this periodic acid-Schiff stain, bar
represents 50 µm.
REFERENCES


J Rheumatol 2010;37:12; doi:10.3899/jrheum.100687
Correction