



A retropharyngeal abscess caused by *Francisella tularensis* with an aberrant internal carotid artery on the same side

*Francisella tularensis*in neden olduđu retrofarengeal apse ve aynı tarafta aberran internal karotis arter

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ABSTRACT

Tularemia is a rare zoonotic disease caused by *Francisella tularensis*, which is a pathogenic spp. of Gram-negative, non-capsular, aerobic coccobacillus. Insect or rodent bite, contact with animals, consuming contaminated animal products, and water may be the routes of transmission. This bacterial infection has a significant place in the otolaryngology practice, as some patients with tularemia have signs and symptoms localized in the head and neck. In this article, we report a 60-year-old female patient with a four-month history of sore throat, dysphagia, right-sided neck mass, and fever. Radiological studies revealed a retropharyngeal abscess with a right-sided aberrant internal carotid artery. To the best of our knowledge, this is the first case of a retropharyngeal abscess caused by *Francisella tularensis* with an aberrant internal carotid artery on the same side.

Keywords: Aberrant carotid artery; *Francisella tularensis*; retropharyngeal abscess.

ÖZ

Tularemia Gram-negatif, kapsülsüz, aerobik bir kokobasilin patojenik suşu olan *Francisella tularensis*in neden olduđu nadir görülen bir zoonozdur. Bulaş yolları böcek veya kemirgen ısırığı, hayvanlarla temas, hayvansal ürünlerin tüketilmesi ve suyla olabilmektedir. Tularemili hastaların bazılarında baş ve boyun yerleşimli bulgu ve belirtiler olduđu için, bu bakteriyel enfeksiyonun kulak, burun, boğaz uygulamasında önemli bir yeri vardır. Bu yazıda, dört aydır boğaz ağrısı, yutma güçlüğü, sağ taraflı boyun kitlesi ve ateş öyküsü olan 60 yaşında bir kadın olgu sunuldu. Radyolojik çalışmalarda, sağ taraflı aberran internal karotis arter ile birlikte retrofarengeal apse izlendi. Bilgimiz dahilinde, bu, aynı tarafta aberran internal karotis arter ile birlikte *Francisella tularensis*in neden olduđu ilk retrofarengeal apse olgusudur.

Anahtar sözcükler: Aberran karotis arter; *Francisella tularensis*; retrofarengeal apse.

Francisella tularensis (*F. tularensis*) is an aerobic, non-capsulated, gram-negative coccobacillus.^[1] This bacteria is one of the most infectious pathogenic agents known, requiring inhalation or inoculation of as few as 10 microorganisms to initiate human infection.^[2,3] Two main types of *F. tularensis* have been described, *F. tularensis tularensis* (type A) which is only found in North America and highly virulent, *F. tularensis holarctica* (type B) which

is less virulent and found in the northern hemisphere.^[4,5] Human infection tends to occur generally in rural areas.^[6] The clinical picture may develop glandular, ulceroglandular, oculoglandular, oropharyngeal, pulmonary and septic forms, depending on the site of transmission.^[5,7] Tularemia is considered a potential threat as a biological weapon because of its virulence and the way of transmission as aerosolized droplets.^[8,9]

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After an incubation period of 3-5 days (range from 1 to 21 days), various clinical manifestations can arise from asymptomatic illness to fulminant toxemia with septic shock, but the most common clinical presentations are fever, sore throat, body aches, coryza, chills and rigor which of all are nonspecific.^[10] Fever and severe throat pain are the main symptoms of the oropharyngeal form (representing up to 12% of cases overall) that presents with pharyngitis or tonsillitis with oral ulcers and unilateral cervical, preauricular and retropharyngeal lymphadenopathy.^[11-14] This form can be acquired by exposure to contaminated water or consumption of poorly cooked meat.^[15] Glandular tularemia involves lymphadenopathy, generally cervical in the deep jugular chain without an identifiable skin lesion and 42% of cases feature high fever associated with bradycardia (pulse-temperature dissociation).^[10,16]

Here, we present a case of tularaemia that caused a retropharyngeal abscess on the same side as an aberrant internal carotid artery.

CASE REPORT

A 60-year-old woman who had type 2 diabetes mellitus and hypertension was admitted to a community hospital with symptoms of fever, chills, sore throat and malaise. Her symptoms persisted despite therapy with several oral antibiotics. After one month, she reported a right-sided upper cervical swelling and dysphagia and received parenteral ceftriaxone 1 gr. three times per day and ciprofloxacin 400 mg two times per day. After parenteral antibiotic therapy, her temperature normalized but cervical swelling and dysphagia persisted.

Approximately two months after her symptoms started, she was transferred to our clinic. Physical examination demonstrated a 4x2 cm, semisolid, fixed right upper cervical mass. On oral examination and fiberoptic upper airway endoscopy a giant bulge was seen extending from the posterolateral nasopharynx to the epiglottis, medializing the right tonsil and arytenoids (Figure 1). Her white blood cell (WBC) count was 7370/mm³ and erythrocyte sedimentation rate (ESR) was 58 mm/h. Serum electrolytes and liver and kidney function tests were normal. Ultrasonography of neck revealed a 40x20 mm, multicystic, necrotic lymphadenopathy in the right cervical jugulodigastric region. Magnetic resonance imaging (MRI) and contrast enhanced computed tomography (CT) identified submandibular and upper cervical necrotic lymphadenopathy (Figure 2) and also a 30x10 mm fluid collection extending in the retropharyngeal space from the level of the second cervical vertebrae (C2) and inferior part of the nasopharynx to the ventral

side of the longus capitis muscle (Figure 3). Contrast-enhanced MRI demonstrated ring-shaped opacification around the collection and these scans were similar to the retropharyngeal abscess. Magnetic resonance and contrast enhanced CT imaging also revealed the abnormal course of the right internal carotid artery (ICA) corresponding to the retropharyngeal space at the level of the nasopharynx and oropharynx; this abnormal course of the ICA had been considered a continuation of the abscess during fiberoptic upper airway endoscopy. (Figure 2, 4). Francisella agglutination test yielded a positive result (1/640).

The patient received doxycycline 100 mg two times per day and streptomycin 1 gr one times per day and enlarged, necrotic cervical lymph nodes were aspirated and aspirates analyzed microbiologically. Since microbiological cultures were negative, the patient was taken to the operating room. During surgery, a multilocular, thick-walled, abscessed lymph node and retropharyngeal abscess was drained. After surgery, all of her symptoms resolved completely in a week. She received doxycycline for three weeks and streptomycin for two weeks. Written informed consent was obtained from the patient to report this case.

DISCUSSION

Consumption of contaminated food and uncontrolled spring water are considered to be the main mode of transmission of *F. tularensis* in Turkey.^[17,18] The oropharyngeal type is more common in Turkey although the ulceroglandular type is the most common type of

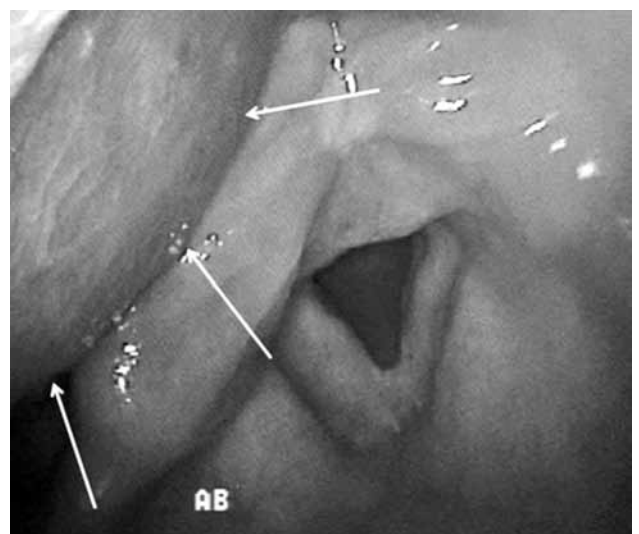


Figure 1. A giant bulge extending from the posterolateral nasopharynx to the epiglottis, medializing the right tonsil and arytenoids.

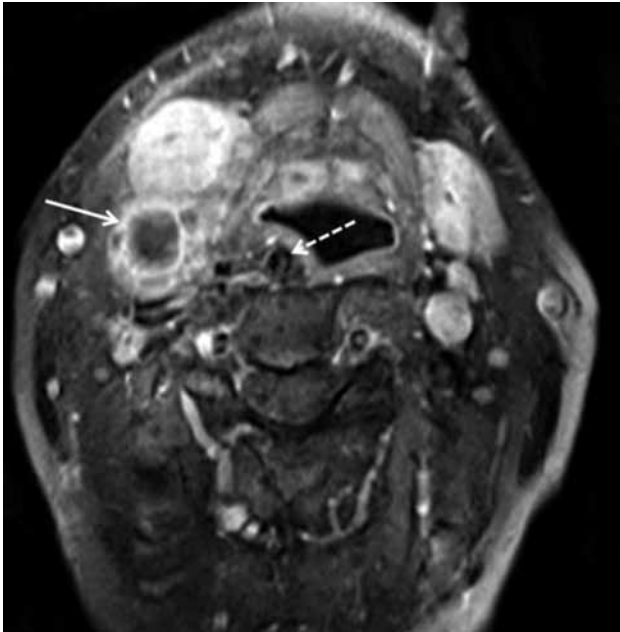


Figure 2. Axial magnetic resonance imaging showing submandibular and upper cervical necrotic lymphadenopathy with aberrant internal carotid artery on the same side.

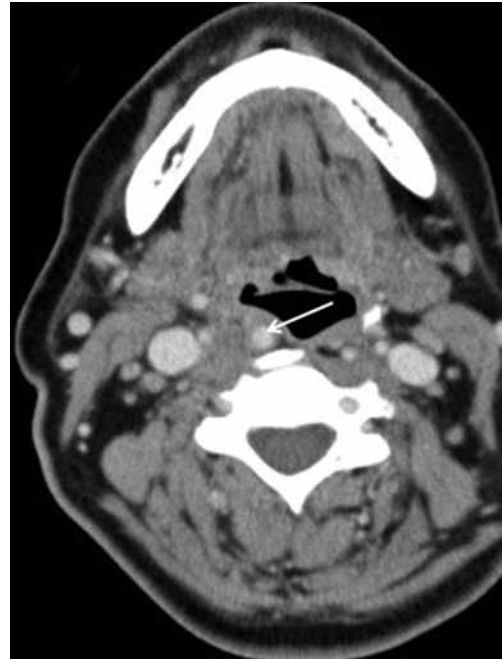


Figure 4. Contrast enhanced computed tomography showing aberrant internal carotid artery.

tularemia in the literature.^[11,17] Approximately 85% of patients have enlarged lymph nodes, especially in the cervical and periauricular region and these enlarged lymph nodes are the only clinical sign in 50% of patients with *F. tularensis*.^[19,20]

Based on duration, cervical lymphadenopathy is classified into three groups; acute lymphadenopathy (less than 2 weeks), subacute lymphadenopathy

(2-6 weeks), chronic lymphadenopathy (more than 6 weeks).^[21] Tuberculosis, Hodgkin disease, non-Hodgkin lymphoma, leukemia, HIV infection, Kikuchi-Fujimoto disease, cat-scratch disease, toxoplasmosis and tularemia are considered in the differential diagnosis of chronic cervical lymphadenopathy.^[22,23] These diseases can mimic each other. Serology, neck and chest imaging, fine needle aspiration biopsy, history of the patient (living in an endemic-zone, history of exposure, presence of skin lesions) and excisional biopsy are used to identify the etiology of chronic cervical lymphadenopathy.

Considering the difficulty of identifying *F. tularensis* by routine microbiological screening protocols, the diagnosis of *F. tularensis* is based upon clinical suspicion. Serology (tube agglutination and microagglutination) is the main process of diagnosing *F. tularensis* microbiologically since the culture and isolation of the bacterium is difficult and hazardous.^[24] Enzyme linked immunosorbent assay (ELISA) is another, safer way of diagnosing this bacteria.^[20] Routine laboratory test results and white blood cell count are also nonspecific in diagnosis and histopathological evaluation does not provide pathognomonic signs for tularemia.^[6]

Abscess formation in a single or multiloculated appearance, enlarged lymph nodes with or without central necrosis and peripheral enhancement following intravenous (IV) contrast media administration were



Figure 3. Sagittal magnetic resonance imaging showing retropharyngeal abscess.

reported as the CT findings of tularemia cases with neck abscess.^[25] Ugur et al.^[6] described the MRI findings of two cases with tularemia, wherein cystic lesions located on the medial part of the sternocleidomastoid muscle with peripheral enhancement and irregular borders consistent with abscess formation were detected on MRI. Therefore, tularemia must be kept in mind in the differential diagnosis of necrotizing lymphadenitis of the neck.

For the treatment of all forms of tularemia, the drug of first choice is streptomycin.^[20] Gentamycin is also another effective antibiotic but macrolides, betalactams, trimoxazole and lincosamides are not preferred.^[21] Ciprofloxacin, doxycycline and chloramphenicol have also been demonstrated to be effective. The therapy must be given at least for 7-14 days. In the presence of a fluctuant abscess or large lymphadenopathy, surgery is needed.^[8,13] Vaccination is recommended only for laboratory personnel who routinely work with this organism since it provides only incomplete protection.^[10]

An aberrant carotid artery is a very rare variation with an incidence of about 5% in the general population.^[27] This variation of ICA (angulation) is divided into two categories: kinking and tortuosity.^[28] The sharp bend of ICA is called kinking and undulation, elongation, redundancy and S-shaped curve are called tortuosity.^[29] Aberrant carotid artery may cause fatal complications during routine otolaryngology procedures such as tonsillectomy, peritonsillar and retropharyngeal abscess drainage, biopsy and pharyngoplasty. In all oral lesions, caution should be taken especially when there is a palpable pulsation or bruit to prevent massive bleeding.

In conclusion, it is not impossible for a patient to have infrequent clinical presentations and rare anatomical variations at the same time as we report in this article. *F. tularensis* may cause, retropharyngeal, peritonsillar and parapharyngeal abscess, and vascular variations must be considered in all oral masses. Contrast enhanced CT and possibly, MRI should be performed before a surgical procedure in such cases.

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