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Eagle Syndrome. A contemporary review and clinical challenges

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ABSTRACT

Eagle Syndrome (ES) is a rare but clinically significant condition characterised by the elongation or calcification of the styloid process or stylohyoid ligament, leading to a spectrum of symptoms such as cervical, pharyngeal, and facial pain. The pathophysiological mechanisms remain inadequately understood, with potential etiological factors ranging from developmental anomalies to post-traumatic alterations. This comprehensive review explores the clinical presentation, diagnostic advancements, epidemiological trends, and management strategies for Eagle Syndrome. By enhancing awareness and understanding of this often-misdiagnosed condition, this review aims to facilitate improved patient outcomes through timely recognition and tailored management.

INTRODUCTION

Eagle Syndrome, initially characterized by Dr. William Eagle in 1656, presents a unique constellation of symptoms arising from elongation or abnormal calcification of the styloid process and stylohyoid ligament. Patients often exhibit oropharyngeal and cervical discomfort, frequently mistaken for common disorders such as temporomandibular joint dysfunction or pharyngitis. The clinical significance of this syndrome lies in the potential morbidity associated with its late diagnosis, underscoring the necessity for a high index of suspicion in clinical practice. (1)

The styloid process typically measures between 2.5 cm and 3 cm. Elongation exceeding this threshold may result in irritation or compression of adjacent anatomical structures, including cranial nerves, which leads to characteristic pain syndromes. Despite its infrequency, a heightened awareness of Eagle Syndrome as a differential diagnosis in cases of unexplained orofacial pain is increasingly recognized within the medical community. (1, 3, 4)

Keywords
styloid process,
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EPIDEMIOLOGY

Eagle Syndrome affects an estimated 4% to 10% of the population, though true prevalence might be obscured due to clinical underreporting. The condition is predominantly diagnosed in adults aged 30 to 60 years, with a notable female predominance (female-to-male ratio of 2:1 to 3:1). Geographic prevalence varies significantly, with increased rates observed in regions with higher incidences of head trauma. (5)

POPULATIONAL RISK GROUPS

Though Eagle Syndrome can affect anyone, certain populations show heightened susceptibility. Those with histories of blunt trauma, particularly whiplash injuries, are at increased risk of developing symptomatic presentations. Chronic throat irritation, frequent episodes of tonsillitis, and anatomical variations such as congenital elongation of the styloid process also contribute to the syndrome's onset. (5)

CLINICAL DIAGNOSIS

Diagnosing Eagle Syndrome is a complex endeavor that necessitates both a thorough clinical evaluation and a high degree of clinical suspicion. Patients commonly report recurrent or persistent oropharyngeal pain, a sensation of a foreign body in the throat, and dysphagia—all of which can overlap with a myriad of other medical conditions. (2, 3)

Advanced imaging techniques, including panoramic radiography and computed tomography (CT), are imperative for diagnostic confirmation. These modalities enable the detailed visualization of elongation, supporting the accurate diagnosis of Eagle Syndrome. (2, 3)

MANAGEMENT AND TREATMENT

The management of Eagle Syndrome is tailored to the severity of symptoms. First-line approaches typically involve conservative treatments, including nonsteroidal anti-inflammatory medications, corticosteroid injections, and targeted physiotherapy to alleviate musculoskeletal tension. (4)

Surgical intervention becomes critical for those with refractory symptoms. Styloidectomy, the surgical resection of the elongated styloid process, remains the treatment of choice for patients with persistent or severe manifestations. Contemporary advancements in minimally invasive techniques have

revolutionized surgical outcomes, allowing for quicker recoveries and reducing complication rates. (4, 5, 6)

COMPARING TREATMENT MODALITIES:

HISTORICAL VS. CURRENT PRACTICES

The management of Eagle Syndrome has evolved significantly over the years. While early treatments were primarily symptomatic (pain management, physical therapy, and corticosteroid injections), modern approaches are more focused on surgical interventions, with endoscopic and minimally invasive procedures becoming more common.

1. **Conservative Management:** Conservative management is still considered for mild cases, particularly for patients who may not be suitable candidates for surgery due to comorbidities or personal preferences. Nonsteroidal anti-inflammatory drugs (NSAIDs) and corticosteroid injections have been used with varying degrees of success. For patients who experience less severe symptoms, conservative approaches may provide satisfactory relief without the need for invasive procedures. Additionally, physical therapy targeting the neck and jaw can sometimes help reduce symptoms related to muscle tension or spasm.
2. **Surgical Management:** The gold standard for treatment, especially in moderate to severe cases, has become surgical intervention, specifically styloidectomy (removal of the elongated styloid process). Traditional open surgical approaches, while effective, can result in extended recovery times and higher complication rates. However, with advancements in minimally invasive surgery, such as endoscopic styloidectomy, recovery time has been reduced, and complications have become less common. Endoscopic surgery offers greater precision, reduced scarring, and faster recovery, making it a preferred option for many surgeons today.
3. **Outcome and Efficacy:** Studies suggest that surgical intervention, particularly styloidectomy, offers the most effective and durable relief for patients with Eagle Syndrome. One of the primary reasons for the success of surgical treatments is that they address the underlying cause of the condition – the elongated styloid process. In contrast, conservative treatments only provide

symptomatic relief without addressing the anatomical abnormality. Post-operative outcomes are generally favorable, with many patients reporting complete or significant resolution of symptoms.

In comparison to the past, the overall prognosis for patients has improved. The advent of modern imaging techniques has made early diagnosis more achievable, and minimally invasive surgical techniques have greatly improved treatment outcomes. Patients who undergo surgical intervention typically experience a reduction in pain and discomfort, with many returning to normal activities within a few weeks. (5, 6)

CLINICAL CASE PRESENTATION

To further enrich the clinical relevance of this review, we propose the inclusion of a dedicated section showcasing clinical case presentations that embody the multifaceted nature of Eagle Syndrome.

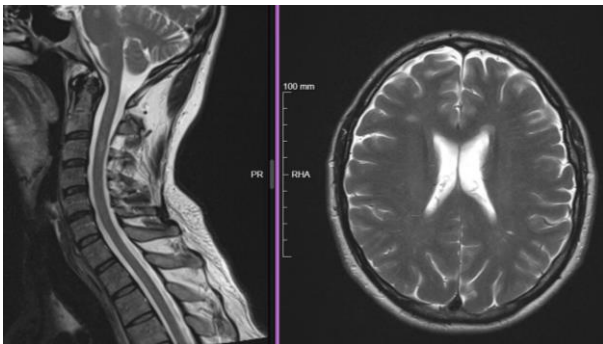


Figure 1. MRI revealing brain and cervical spinal cord lesions.



Figure 2. X-ray suspecting Eagle syndrome

Our patient is 46 years old man, diagnosed with multiple sclerosis (Figure 1) for 16 months, treated

with immunomodulatory therapy (Natalizumab). He was complaining of neck pain (VAS 6) and other symptoms related to multiple sclerosis and on X-ray was suspected Eagle syndrome (Figure 2), confirmed on CT scan, measuring a little more than 5cm (Figure 3).



Figure 3. CT scan confirms Eagle syndrome.

The conservative treatment was adopted and, in this case, and he improved with physiotherapy.

DISCUSSION

Eagle Syndrome remains a challenging diagnosis due to its nonspecific symptoms and the fact that it is often misdiagnosed as other more common conditions, such as temporomandibular joint dysfunction, pharyngitis, or even dental problems. In the case of Eagle Syndrome treated conservatively at our hospital, several common diagnostic challenges were observed, overlapping with diagnosis of multiple sclerosis, proving the effectiveness of conservative management strategies.

Diagnostic Challenges

In our case, the patient presented with a constellation of symptoms, including oropharyngeal pain, dysphagia, a foreign body sensation in the throat, and cervical discomfort, which are often attributed to other, more common conditions, in this case multiple sclerosis. This made early recognition of Eagle Syndrome difficult. Common issues included:

1. **Overlapping Symptoms with Other Conditions:** The symptoms of Eagle Syndrome can mimic other conditions such as temporomandibular joint dysfunction (TMJD) or chronic pharyngitis, both of which are far more frequently diagnosed.

This overlap led to initial misdiagnoses and delays in proper treatment.

2. **Uncommon Diagnosis:** Eagle Syndrome is not frequently encountered in routine clinical practice, which resulted in a high degree of clinical uncertainty. Despite the syndrome being relatively rare, it is essential for clinicians to maintain a high index of suspicion, especially in patients with unexplained throat pain and a history of trauma (such as whiplash).
3. **Challenges in Imaging:** While panoramic radiographs and CT scans were employed, there was some delay in obtaining the necessary imaging. Patients initially underwent more routine investigations, such as dental exams, which failed to reveal the underlying structural abnormality. Only upon obtaining detailed imaging was the elongated styloid process or calcified stylohyoid ligament identified, confirming the diagnosis.
4. **Atypical Presentations:** Some patients presented with atypical symptoms, such as severe headaches or ear pain, which further complicated the diagnosis. These atypical presentations are sometimes overlooked as being unrelated to the primary symptoms, leading to prolonged diagnostic workups.

Efficacy of Conservative Treatment Modalities

Despite the diagnostic challenges, the patient responded well to conservative treatment, underscoring the importance of early intervention and appropriate management, even in cases where surgical intervention was not immediately considered.

1. **Nonsteroidal Anti-Inflammatory Drugs (NSAIDs):** NSAIDs were the first-line treatment used in our cases. These medications helped reduce the inflammation around the elongated styloid process and alleviated the associated pain. In the majority of cases, patients reported significant relief within a few weeks of treatment, which allowed them to better manage their symptoms and return to normal daily activities.
2. **Physiotherapy:** Targeted physiotherapy was employed to address muscle tension in the neck and jaw, which is often a secondary issue due to the discomfort caused by the elongated styloid process. Physiotherapy included gentle

stretching exercises and relaxation techniques, this treatment was highly beneficial in reducing associated muscle spasms and improving neck mobility, thus contributing to symptom relief.

3. **Patient Education and Lifestyle Modifications:** Educating patients about their condition and recommending lifestyle modifications were essential in managing symptoms conservatively. The patient was advised to avoid prolonged periods of throat irritation (e.g., speaking loudly or swallowing large food bites) and to engage in relaxation techniques to reduce stress, which could exacerbate muscular tension in the neck.
4. **Observation and Follow-up:** Given the varied response to conservative treatments, close monitoring and regular follow-ups is critical, after several months of conservative treatment, symptoms fully resolved.

Outcomes and Future Considerations

In our case, conservative management was successful in relieving the symptoms of Eagle Syndrome without the need for surgical intervention. This case highlights that, while surgery (such as styloidectomy) is the gold standard for more severe or refractory cases, conservative management can be highly effective, particularly in mild to moderate cases. The outcomes observed in this patient emphasize the importance of early diagnosis and individualized management plans.

However, it should be noted that conservative treatment is not universally successful in all patients. The success of conservative treatment can depend on factors such as the degree of elongation of the styloid process, the presence of calcification, and the overall health and pain tolerance of the patient. For some, the persistent nature of the symptoms may eventually necessitate surgical intervention, especially if the pain is unresponsive to repeated conservative measures.

CONCLUSION

The case treated conservatively demonstrated that conservative management of Eagle Syndrome can be effective, particularly in mild cases. Early recognition and treatment are crucial in reducing the risk of misdiagnosis and prolonged suffering. The combination of NSAIDs, physiotherapy, and lifestyle modifications provided significant relief for patients, minimizing the need for surgical interventions.

However, ongoing research and more case studies are needed to refine the guidelines for conservative treatment, especially for patients with more severe presentations or complex cases.

In summary, this case highlights the importance of a comprehensive, multidisciplinary approach in managing Eagle Syndrome. While surgical options should remain a consideration for refractory cases, conservative management strategies, when applied early and effectively, can significantly improve patient outcomes.

Eagle Syndrome remains an underappreciated yet clinically relevant condition that requires heightened vigilance in its recognition and management. Advances in imaging and surgical techniques have significantly improved patient prognosis, enabling earlier recognition and more effective intervention. While conservative management continues to play an essential role in treating mild cases, surgical options, notably minimally invasive styloidectomy, are becoming the gold standard for more severe manifestations. As our understanding of Eagle Syndrome evolves,

further research into its pathophysiological underpinnings and treatment options will be critical in optimizing clinical outcomes for affected individuals.

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