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Fever and acquired torticollis in hospitalized children

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Abstract

Acute torticollis due to non-traumatic atlanto-axial subluxation (AAS) is often seen in children presenting with inflammatory conditions of the upper respiratory tract and the neck. Grisel's syndrome is the eponym given to this condition. These patients may present earlier in the disease process without evident subluxation. Thus, early recognition of the condition with prompt commencement of appropriate conservative treatment could halt the progression into Grisel's syndrome. The purpose of this study is to address the importance of early recognition of inflammatory torticollis that can be treated successfully by conservative methods.

A retrospective review was made of the case files and radiological investigations of 13 children with fever and torticollis who were treated in the neurosurgery unit of Hamad General Hospital in Qatar, over a two-year period from July 1996 to July 1998. The children were aged between three and 12 years with a male to female ratio of 3:1. All patients arrived at the hospital within 48 hours of onset of torticollis and almost all had manifestations of upper respiratory tract or head and neck infections. Radiological examination by cervical spine X-rays, computerized tomography (CT) or magnetic resonance images (MRI) revealed that only three cases out of 13 had AAS. All patients underwent conservative treatment that included rest, neck collar, simple analgesics and antibiotics, where appropriate. A muscle relaxant was used in nine cases and Halter traction was applied to the three with AAS. All patients responded well to treatment and none required surgical intervention for AAS.

We conclude that the majority of children presenting acutely with inflammatory torticollis have rotational deformity only without AAS. Progression to the latter, i.e. Grisel's syndrome, may be aborted should the diagnosis be made early and conservative treatment initiated in time. On the other hand, delay in diagnosis would deprive these children an opportunity of receiving effective conservative treatment.

Key words: Torticollis; Fever; Child

Introduction

Torticollis, a descriptive term implying rotational deformity of the cervical spine with secondary turning or tilting of the head can result from diverse causes, the exact pathogeneses of which are not completely understood. This is reflected in the various terminologies, treatment modalities and treatment outcomes reported in the literature. Frequently, the rotational deformity of the spine is associated with subluxation of the atlas on the axis. Although first described by Bell in 1830, non-traumatic atlanto-axial subluxation associated with inflammatory processes in the neck has been given the eponymous name of Grisel's syndrome.[1] The infection may be overt or covert, local to the cervical region or metastatic in the form of cervical adenitis.

Some patients present with fever and torticollis without atlanto-axial subluxation and these patients with only a rotational deformity, strictly speaking, should not be classified together with those with subluxation. This distinction, which is often ignored, explains why treatment protocols and prognostications have been so varied in the literature.

Fielding *et al.* classified atlanto-axial rotatory deformity into four distinct groups, that are still valid.[2,3] In type I, which is by far the commonest, are patients with rotatory deformity without anterior displacement of the atlas or with a displacement of 3 mm or less. In these patients the transverse ligaments remain intact. While it is valid to consider patients with intact transverse ligaments as a group from the point of view of pathogenesis, the clinical course, treatment modality and prognosis depend to a large extent on the presence or absence of subluxation.

The presence of atlanto-axial subluxation is not always easy to demonstrate in the child. Trans-oral and lateral cervical spine radiographs should contribute significantly to this diagnosis but are rarely achieved to a standard quality in the uncooperative

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child with torticollis. Spiral CT-scans with 3-D reconstruction have been shown to add to the resolution.[4]

In this retrospective study we have analysed the clinical presentation, plain radiographs, CT scans and MRI scans of children with torticollis, in association with fever, to determine the presence or otherwise of atlanto-axial subluxation. Treatment method and outcome were compared in the two groups.

Materials and methods

The case records of 13 children admitted to the Hamad General Hospital with fever and torticollis between July 1996 and July 1998 were retrospectively reviewed. Fever was defined as a temperature of 37.5°C or above at the time of admission. Patients with a clear history of fever and a documented temperature of 37.5°C or above from primary health care facilities were also included. All patients who developed fever after hospitalization were excluded.

Clinical, radiological and laboratory data were collated and analysed for possible aetiology and related pathogenesis. The type, timing and indication for radiological examination were noted and the contribution of these data to the modality of treatment assessed. Patients were broadly categorized into those with, or without, subluxation. Treatment outcome and duration of hospitalization were then compared in the two groups. Follow-up was continued for a minimum period of six months. Neck deformity, pain resolution and movement limitations were the end point criteria for outcome.

Results

A total of 13 children aged between three and 12 years entered the study. All patients arrived in hospital within 48 hours of onset of torticollis. The male to female ratio was 3:1 (Table I). Temperatures ranged from 37.6°C to 40°C. Pain of sudden onset was the most common presentation and the neck was more frequently turned to the left although this was not significant. Evidence of upper respiratory tract infection was present in the form of cough, sore throat and pharyngitis (Table II). Seven patients had palpable cervical lymph nodes and the tonsils were enlarged or inflamed in four (Table III).

Two patients had an antecedent history of trauma. In these patients, the onset of torticollis was immediate, both had a temperature below 38°C (37.6°C and 37.9°C), and one also had pharyngitis and enlarged tonsils (Table I).

Cervical radiographs were obtained in all patients in the anterior-posterior and lateral views. Additional trans-oral views were possible only in six patients. Apart from the tilting of the neck and loss of normal curvature, no obvious abnormalities were noted in the cervical radiographs of nine patients. Atlanto-axial subluxation was noted in two patients and pre-vertebral soft tissue swelling in two (Table

TABLE I
SUMMARY OF CASES

Serial No.	Age (yrs.)	Sex	National	Side of head twist	Type	Mode of onset	Predominant symptoms	Predominant signs	Radiology (Plain Film)	Treatment	Hospital stay (days)
1	3	M	Jor	L	Painful	Gradual	Fever diarrhoea Vomiting Temp. 39.5°C	Cervical lymphadenopathy	Normal	I	6
2	3	M	Qat	L	Painful	Sudden	Fever, cough, sore throat. Temp. 37.7°C	Lymphadenitis Pharyngitis	Normal	I + B	4
3	4	M	Qat	R	Painful	Sudden	Fever, ear-ache, sore throat. Temp. 38.5°C	Pharyngitis Otitis	Normal	I + A	5
4	4½	F	Qat	L	Painful	Gradual	Fever, vomiting, neck swelling	Lymphadenitis Tonsillitis Pharyngitis	Pre-vertebral soft tissue swelling	I + A + B	4
5	5	M	Qat	L	Painful	Sudden	Fever, cough, sore throat. Temp. 38.0°C	Pharyngitis	Normal	I + A + B	4
6	5	M	Qat	R	Painless	Gradual	Fever, cough Temp. 39.5°C	Lymphadenopathy Chest - consolidation	Normal	I + A	2
7	6	M	Kuw	L	Painful	Gradual	Fever, dental - abscess Temp. 38.3°C	Cervical lymphadenopathy	Normal	I + A + B	8
8	6	M	Egy	R	Painful	Sudden	Fever, cough sore throat Temp. 39.6°C	Pharyngitis lymphadenopathy	Prevertebral soft tissue - swelling	I + A + B	3
9	6	M	Qat	R	Painful	Sudden	Fever, sore throat Temp. 39.8°C	Tonsillitis lymphadenitis	Normal	I + A + B Traction	12
10	7	F	Pal	L	Painful	Sudden	Fever, cough trauma Temp. 37.9°C	Pharyngitis enlarged tonsils	Normal	I + A + B	4
11	7	M	Qat	R	Painful	Sudden	Fever, Temp. 39.8°C	–	Normal	I + B	3
12	8	M	Qat	R	Painful	Sudden	Fever, sore throat Temp. 38.3°C	Enlarged tonsils	Atlanto-axial sub-luxation	I + A + B Traction	5
13	12	F	Qat	L	Painful	Sudden	Fever, trauma Temp. 37.6°C	–	Atlanto-axial sub-luxation	I + B Traction	10

I = analgesic and collar; A = antibiotics; B = benzodiazepine

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TABLE II
CLINICAL FEATURES: SYMPTOMS

Presenting symptoms	No.	%
Fever	13	100
Neck pain	12	92.3
Onset of pain		
– Sudden	9	75
– Gradual	3	25
Side of Torsion		
– Left	7	53.8
– Right	6	46.2
Sore throat	6	46.2
Cough	5	38.5
Vomiting	2	15.4

IV). Eight patients were subjected to CT either because of suspicious plain radiographs (five) or delayed resolution of pain and deformity (three). Three cases of atlanto-axial subluxation were identified (Figure 1). One of these had been missed by plain radiographic analysis. Five children had MRI of the cervical region (Figure 2). There was obvious pre-vertebral soft tissue swelling in three and C1-C2 rotation abnormality in two (Table IV).

Treatment was conservative in all patients with simple analgesic, rest and cervical collar (Table I). Low dose diazepam was additionally used in nine patients and antibiotics in seven. The three patients with radiological evidence of atlanto-axial subluxation were managed with Halter traction and in two, reduction was achieved in the first 48 hours. In the third patient, traction was started on the fourth day of admission because of a delay in diagnosis and was needed for six days. None of the patients required surgical intervention. The duration of hospital stay was less than three days in three patients and three patients stayed for over one week (Table I). Six patients continued to have some limitation of cervical movement at discharge but none had any significant residual deformity. All patients were discharged on soft cervical collar, for at least three weeks. At the end of three months, all patients demonstrated full resolution of pain and deformity, and a full range of neck movement. Follow-up CT scan in the three patients with subluxation showed no persisting abnormality.

TABLE III
CLINICAL FEATURES: SIGNS

Presenting signs	No.	%
Limitation of neck movt.		
mild	3	23.1
moderate	7	53.8
severe	3	23.1
Temperature °C		
37.5–38.4	6	46.2
38.5–39.4	1	7.7
39.5–40.0	6	46.2
Pharyngitis	6	46.2
Tonsillitis	4	30.8
Cervical lymphadenopathy	7	53.8
Trauma	2	15.4
Others	3	23.1

TABLE IV
RADIOLOGICAL FEATURES

Plain cervical radiographs	
Normal	9
Subluxation	2
Prevertebral soft tissue	2
Cervical CT scan	
Not done	4
Normal	5
Subluxation	3
Cervical MRI scan	
Not done	8
Soft tissue swelling	3
Rotation C1–C2	2

Discussion

The pathogenesis of torticollis secondary to inflammatory processes has remained a matter of some controversy. It has long been held that infection causes periligamentous inflammation, which results in laxity and consequently subluxation of the atlanto-axial joint.[5,6] There is some anatomical evidence[7] to support the view that ligamentous laxity may be secondary to peri-ligamentous inflammation. Recently however, Welinder *et al.*[8] argued, based on serial CT scans in a six-year-old boy with Grisel's syndrome, that the pathogenesis may be more

FIG. 1
Lateral cervical radiograph showing a marked widening of the pre-vertebral soft tissue in the upper cervical region. The normal cervical lordosis is lost.

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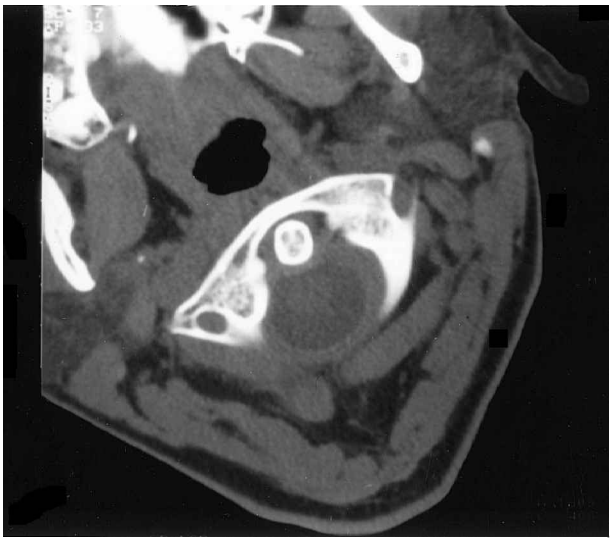

FIG. 2

C1-C2 axial CT scan demonstrating type 1 rotatory fixation. The dens is slightly eccentric in position. The transverse ligament is intact and there is no anterior displacement of the atlas. Note the neck twist to the right.

FIG. 3

Saggital T2 weighted MR scan showing pre-vertebral collection.

related to a distension of the ligament rather than laxity. About 69.2 per cent of children in our study developed rotational deformity associated with systemic inflammatory process but without any evidence of ligamentous inflammatory collection on radiological examination. The early rotational deformity without displacement is most probably due to muscle spasm, which as most authors now believe is a protective reflex rather than the cause of the displacement as earlier suggested by Grisel.[9] The remaining four had some evidence of soft-tissue swelling and in three this was associated with subluxation. It would seem that the progression to subluxation is related to a progression of the inflammatory process. It is possible that early radiological evaluation as well as early antibiotic therapy may have prevented the appearance of significant soft tissue inflammatory collection in the first group, thus aborting the progression of the process to subluxation. At what point subluxation sets in during the course of the inflammatory process however remains to be evaluated by studies with serial MRI or CT scans.

Upper respiratory tract infection remains the commonest cause of Grisel's syndrome, and in many of these patients cervical lymphadenitis precedes the onset of torticollis suggesting a causal relationship. However, torticollis may occur without any evidence of palpable or inflamed cervical lymph nodes, as seen in six of our patients. It is known that any inflammatory process in the upper cervical region, including trauma, which irritates the cervical muscles, nerves or vertebrae could produce reflex spasm resulting in torticollis.[10] While tonsillitis and retro-pharyngitis may cause this directly, more distant infections in the ear, chest or indeed any other site in the body may lead to cervical adenitis and thus torticollis. In two patients, febrile illness was associated with trauma. The fever might have been reactive to the trauma in one case but in the second it was antecedent to trauma and the patient had evidence of upper respiratory tract infection. There was no clear relationship between the degree of pyrexia and the extent of the subluxation.

Trans-oral views demonstrating the atlanto-axial joint can provide excellent information in rotational deformity but unfortunately is not easy in children with painful torticollis. In these patients, a CT scan with or without three-dimensional reconstruction should be considered the examination of choice, especially when evaluating for subluxation.[4] However, in this setting, it should be noted that dynamic CT examination, i.e. CT scanning with the head rotated to each side is important to differentiate between pathological and physiological subluxation. In one of our patients, atlanto-axial subluxation was missed on plain cervical radiograph and this caused a delay in diagnosis and in instituting the appropriate treatment. CT scan was only carried out following a failure of clinical improvement 72 hours after admission. This could have been avoided if a policy of CT scan on initial evaluation was used. Fortunately, in this case no adverse consequences resulted from the delay. However, as shown by many workers,[11,12] delays in initiating traction can result in an increased need for surgical intervention. Early CT scanning in anticipating this problem may reduce the duration of hospital stay and actually reduce costs.

In patients and torticollis, without CT evidence of subluxation, simple measures alone as advocated by Boiten *et al.*[13] are sufficient to achieve resolution of pain and deformity. Paracetamol, soft cervical collar and bed rest in the early, very painful period was sufficient in most patients. There is, however, no need to restrict the child to bed once pain settles and indeed this may be difficult in the active child and is of no benefit. Antibiotics may be necessary, especially with established tonsillar, dental, chest and ear infections.

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If spasm persists after the first 24 hours, we found that adding 1–2 mg of diazepam twice or thrice daily improved response. Traction was used only for patients with obvious subluxation. The decision whether or not to use traction should be made on admission. The reported difficulties with the management of torticollis result from delays in estalishing adequate treatment in the form of early traction when indicated.[11] Phillips and Hensinger[12] showed that successful reduction and the time it took to achieve it were both related to the duration of symptoms as was shown in one of our patients. The presence or otherwise of subluxation must therefore be established early and this is best achieved if CT scan is done routinely on admission in patients with torticollis.

If appropriate treatment is adopted, most patients improve rapidly. Pain resolution usually occurs before recovery of the limitation of movement. We did not wait for recovery of full range of neck movement before discharge. Once pain subsides and the child can voluntarily put the neck in the neutral position, we allow them home. When reviewed after one month all our patients showed full recovery and this was sustained at six months.

Conclusion

Torticollis resulting from inflammatory processes in the neck is not always associated with atlanto-axial subluxation. In the majority of cases, only a rotational deformity may occur in the early phase, although this can quickly progress to subluxation. Treatment modality, outcome and prognosis depend on the presence or absence of subluxation and this should be determined early with CT scans. Patients without subluxation should therefore not be classified as Grisel's syndrome, but should be regarded as inflammatory torticollis. Their management, however, must anticipate a rapid progression to Grisel's syndrome. For that reason, we emphasize that physicians and surgeons treating such cases, be aware of the condition with a view to establishing diagnosis at an early stage. Delay in diagnosis is detrimental in this context as these children would otherwise miss an opportunity for receiving a simple but effective conservative treatment modality.

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