A RARE CAUSE OF ACUTE SCROTAL PAIN IN PAEDIATRIC PATIENTS; A QUESTIONNAIRE SURVEY AND LITERATURE REVIEW

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Submitted 4/11/2017; accepted 31/12/2017

ABSTRACT

Background: Acute scrotal pain (ASP) in children is a urological emergency, which usually warrants scrotal exploration. In the paediatric population, Henoch Schonlen Purpura (HSP) can occasionally present with acute scrotal pain and oedema, which may be misdiagnosed with testicular torsion. We presented two cases encountered in our own practice; we also reviewed the literature and assessed awareness of the link between HSP and ASP amongst urologists.

Subject and Methods: A questionnaire consisting of 9 questions was sent to 105 randomly selected urologists practicing in nine different countries including the UK. All of the urologists were currently employed in institutes accredited for training.

Results: Response rate was 62% with 65 completed returns from urologist of previous levels. Only 25 (38%) were previously aware of the HSP-ASP relationship. In those who were aware of the HSP-ASP link, (28%) would still favour immediate scrotal exploration; while the remainder would perform a Doppler ultrasound scan before deciding. Overall, 91% felt that the HSP-ASP relationship was not well known and 82% considered this an important issue that the practicing urologist should be aware of.

Conclusions: Despite variations in practice with regards to the management of ASP, the relationship between HSP and ASP is not well known. Though the mainstay of the management of patients with ASP is immediate scrotal exploration, we consider that practicing urologists need to be made aware of the link between HSP and ASP.


Keywords: Scrotum, Purpura, Schoenlein-Henoch, Spermatic Cord Torsion, Orchiopexy.

A urological emergency with majority of patients undergoing an immediate scrotal exploration to exclude testicular torsion. The differential diagnosis for ASP includes testicular torsion, torsion of the testicular appendix, infection and trauma. However, in rare cases, ASP in young children may be encountered as the first presentation of Henoch-Schonlen Purpura (HSP), a systemic vasculitis affecting multiple organ systems in the body1. We present our experience with two such cases that were encountered in recent months within the Department of Urology and Department of Child Health at East Lancashire Hospitals in Blackburn, a questionnaire based assessment of the awareness of the link between HSP and ASP amongst urologists, as well as a review of related literature.

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CASE PRESENTATION
Two boys (patient A and B) aged 5 and 6 years respectively presented acutely on two separate occasions to the Department of Urology with the primary complaint of unilateral ASP. On initial assessment, both patients were noted to have a preceding history of fever, arthralgia (non-specific joint pain) and rash for few hours prior to the onset of ASP. A general clinical examination confirmed the presence of a low grade pyrexia, petechiae and palpable purpuric rash over the lower extremities. Local examination of patient A revealed a very tender, indurated and erythematous right hemi-scrotum. The testicle itself was in a normal vertical lay with a positive cremasteric reflex. While in patient B the testis was similarly extremely tender, hot and swollen. Slightly oblique in lay with an absent cremasteric reflex. In both patients the pathology was limited to the scrotum and not extending into the cord. Inguinal examination was normal. These examination findings were suggestive of acute testicular torsion and, following detailed discussions with the guardians; arrangements were made for an immediate scrotal exploration, despite consultation with the pediatrics team and confirming the diagnosis of HSP.

During scrotal exploration, there was no evidence of testicular torsion, but both patients had a thickened and edematous scrotal wall, with adherent tunica vaginalis and non-torted but inflamed testis. On that basis, a formal testis fixation (orchidepexy) was not performed. The scrotum was closed and both patients were observed overnight and allowed home the next morning. The rest of the post-operative recovery was uneventful with no evidence of subsequent loss of testis.

PATIENTS AND METHODS
A questionnaire consisting of nine questions was sent to 105 randomly selected urologists practicing in nine different countries including the UK (Figure 1). All of the urologists were currently employed in institutes accredited for training.

RESULTS
The response rate was 62% with 65 completed questionnaire returned (Figure 2).
Of these, 22 (33%) were qualified consultants, while the rest were urologists in training (Figure 3).

![Figure 3: Urologists Level](image)

Only 25 (38%) were previously aware of the HSP-ASP relationship (Figure 4).

![Figure 4: Awareness](image)

Of these, 16 (24%) urologists had known about this having read about it, 5 (8%) had personally encountered a case previously and 4 (6%) knew from other sources (Figure 5). The majority (92%) would not routinely remember to look for other non-urological manifestations of HSP.

![Figure 5: Source of Awareness](image)

In those who were aware of the HSP-ASP link, (28%) would still favour immediate scrotal exploration; while the remainder (72%) would perform a Doppler ultrasound scan before deciding (Figure 6).

![Figure 5: In those Aware](image)

Overall, 91% felt that the HSP-ASP relationship was not well known to them (Figure 7) and 82% considered this an important issue that the practicing urologist should be aware of.
DISCUSSION

Henoch Schonlen Purpura, is a vasculitic syndrome of small blood vessels, characterised by the deposition of immunoglobulin A (IgA)-containing immune complexes in the walls of small vessels (arterioles, capillaries and venules). It is the most common cause of non-thrombocytopenic purpura in children\(^1\), with a reported annual incidence between 10 and 30 cases per 100,000 children <17 years based on hospital and overall population estimates. The mean age of presentation is six years with most cases occurring in children <10 years of age\(^2\). Purpura or petechiae (mandatory) with lower limb predominance and at least one of the four following criteria (abdominal pain, arthritis or arthralgia, renal involvement and histopathology confirming typical leucocytoclastic vasculitis with predominant IgA deposit or proliferative glomerulonephritis with predominant IgA deposit) are required for the definition of HSP as defined by the Paediatric Rheumatology European Society\(^3\). Management comprises of supportive treatment, including analgesics, as the majority of cases resolves without permanent consequences, though in a minority, serious gastrointestinal and renal complications may occur. Involvement of the male genitalia as the initial manifestation of HSP is unusual and the diagnosis can easily be missed\(^4\). Acute scrotal pain has been described in between 2 and 38 percent of patients with HSP, with the first case reported by Allen et al in 1960\(^5\). Scrotal involvement in HSP mimicking testicular torsion is a self-limiting, benign disease and, if required, will respond to corticosteroid and/or antibiotic therapy\(^4\).

An acute scrotum is defined as an acute painful swelling of the scrotum or its contents, which is accompanied by local signs or general symptoms\(^6\). Aetiological factors for the acute scrotum include, but are not limited to, epididymitis, abscess, tumour, torsion of testicular appendage, acute idiopathic scrotal oedema and torsion of the spermatic cord. The latter is of the utmost concern since it requires immediate surgical correction to minimise the risk of testicular loss due to ischaemia. In clinical practice, extreme tenderness over the affected area hampers adequate scrotal examination in children and often the diagnosis is difficult to establish simply on clinical grounds\(^7\). Thus, most patients undergo exploration\(^8\). Some authors have advocated the use of ultrasound study as an adjunct to aid the diagnosis, but this is user-dependent and may miss acute testicular torsion. An experienced sonographer may be able to distinguish between findings of acute testicular torsion and ASP due to HSP, the sonographic features included swollen, hypoechoic
testes with hyperemia, which are suggestive of orchitis due to underlying pathology. These findings may cause an earlier diagnosis of HSP from other scrotal diseases. Others have used radioisotope scans in diagnosis of ASP in children, but this may not be suitable due to concerns about availability.

A high index of suspicion is required to identify those rare cases of ASP secondary to HSP and attending doctors must remember searching for other associated symptoms and signs of HSP in order to help in avoiding unnecessary scrotal exploration. A note of caution is appropriate, as three cases of torsion of a testicle or its appendix have been identified in patients with HSP.

We concluded that despite variations in practice with regards to the management of ASP, the relationship between HSP and ASP is not well known. Acute scrotal pain can still be a manifestation of HSP, and both urologists and paediatricians need to be aware of this relationship. While the majority of children presenting with ASP will still warrant urgent scrotal exploration, increased vigilance, and collaborative multi-institutional efforts will help to identify characteristics and investigations which may help in reducing unnecessary surgical intervention.

AKNOWLEDGMENT

The authors reported no conflict of interest and no funding was received for this work.

REFERENCES


ثوحتة

نَاظَرَتْكِ دَكْطَمَانَ يِنَيْنَا هِيْلَكا يا لَنَضْرِ لَ دَقْفِ زَارُوكَ، ثِيْشِكِشْكِكَنَا جُوَرَيْ نَغْوَشِيْنَ،
دِيارَكَنا ثِيْزَانَيْنَا ذِ جَهِينِ يَأْوَدْرُيْ ثُكْرَى

ثِيْشِكِسْكِةُ:٢٥ يَنْيْنَا دَكْطَمَانَ يا هِيْلَكا لَنَضْرِ لَ دَقْفِ زَارُوكَ، ثِيْشِكِشْكِكَنَا جُوَرَيْ نَغْوَشِيْنَ،
مَيْزِيَُّّ، كَوْ دَلْطِيْنَ بِهِيْتَةَ دِيَارَكَنَّ. لَ دَقْفِ زَارُوكَ بَاتِكَ هِينُوشُ شَوْلَنَ بُورُوُرَا (هُشْب) بِهِيْتَةَ
نِيَاسِينَ يَنْيْكَ ذِ دِيَارَكَنَا دَكْطَمَانَ يَا هِيْلَكا لَنَضْرِ، نَأَوَأَ بَاتِكَ بِهِيْتَةَ نِيَايَنَ خَارِبوُنَا هِيْلَكَ.
نَقْطُ شُلوْطَكَرِنَا هَاتَةُ ثِيْشِكِشْكَيْكَ كَرِنَ بَوْ دُوُو جَوْرَا لَ كَاَرِ مَا بِيْ ثِيْشِكِسْكِثَا، دِتاَتِ دِيَارَكَنَا
بَنَجَيْيِنَّ ثِيْزَانَيْنَا كَوْ دَتْرِيْدَيْنَيْنَا بَنِيْنَا دَكْطَمَانَ يَا هِيْلَكا لَنَضْرِ، وَ هِينُوشُ شَوْلَنَ بُورُوُرَا لَ
لَاَيِيْ نُوْدَارَيْنَ جَوْبَارَيْنَ مَيْزِيَ.

رِيْكُوْنَةَ ثُكْرَيْكِيْنَ:٩ ثُرْسيَرَا هَاتَنَ فَرَيْكَنَ بَوْ ١٠٥ نُوْدَارَيْنَ جَوْبَارَيْنَ مَيْزِيَ نَوْتَيْنَ هَاتِيْنَ
هُتَلْبَارَيْنَ بَثِيْكُوْكَ يَنْ ٩ وَدَلْطَايَا كَارِ دَكْتَانَ وَ دَلْطِيْنَ وَاَلَّيَا بِرُيْنَايَا بَوْ زَانَيْنَ نَقْفُ نُوْدَارَيْنَ هَاتَ
بُونَةُ ثِيْنَا نَخَذَارَا هِمَايَا بَاَرُدَا كَارَ كَرِيْ لَ دُقْزَتَهُنَّ بَاوَرْتَيْ ثُكْرَيْ بَوْ رَاهِيْنَايَا دَضْرَوُضُيْنَ
طَيْ شُلوْطَكَرِنَا دَا.
A RARE CAUSE OF ACUTE SCROTAL PAIN IN PAEDIATRIC PATIENTS

الخلاصة

سبب نادر لألام الخصية الحاد لدى الأطفال؛ تقديم حالة مرضية، تقييم استبيان واستعراض المراجع

الخلفية والأهداف:

ألم الصفن الحاد في الأطفال هو حالة طارئة في جراحة المسالك البولية، والتي عادةً ما تستدعي إستكشاف الصفن. في الأطفال يمكن أن يظهر هينوش شونلن بوربورا (ه شب) في بعض الأحيان كحالة ألم الصفن الحاد مع وذمة، والتي قد تشخص خاطأ على أنها التواء الخصية. قدنا في هذا البحث حالتين واجهتا في ممارستنا الطبية؛ مع استعراض المراجع وتقديم الوعي بالصلة بين ألم الصفن الحاد و هينوش شونلن بوربورا لدى أطباء المسالك البولية.

طرق البحث:

تم إرسال استبيان مكون من 9 أسئلة إلى 105 من أطباء المسالك البولية الذين تم اختيارهم بشكل عشوائي والذين يعملون في تسعة بلدان مختلفة من بينها المملكة المتحدة علما أن جميع هولاء الأطباء عملوا في مؤسسات معتمدة للتدريب أثناء إجراء الاستبيان.

النتائج:

كان معدل الاستجابة للاستبيان %62 حيث تم الحصول على 65 إجابة كاملة للإستبيان من أطباء المسالك البولية أعلاه. بين أن %25 (37) طبيبا كانوا على علم مسبق للعلاقة بين هينوش شونلن بوربورا و ألم الصفن الحاد، وبين أولئك الذين كانوا على بيئة هذا الارتباط بين %25 منهم كانوا يفضلون الاستكشاف الصفني الفوري، في حين أن باقي الأطباء فضلا إجراء فحص الموجات فوق الصوتية (دوبلاي) قبل اتخاذ القرار. عموماً بين أن %91 من الأطباء الذين استجابوا للإستبيان شعروا بأن العلاقة بين هاتين الحالتين لم تكن معروفة جيدا وأن %82 من هؤلاء الأطباء يعتبرون أن هكذا حالة هي قضية هامة على طبيب المسالك البولية أن يكون على بيئة منها.

الاستنتاجات:

على الرغم من الاختلافات في الممارسات فيما يتعلق بعلاج ألم الصفن الحاد فإن علاقة هذه الحالة مع هينوش شونلن بوربورا ليست معروفة جيداً، وعلى الرغم من أن العلاج الأساسي للمرضى الذين يعانون من ألم الصفن الحاد هو الاستكشاف الصفني الفوري تعتقد أنه يتعين على أطباء المسالك البولية أن يكونوا على دراية وعلم بالصلة بين هاتين الحالتين.