

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

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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.

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

Acute scrotal pain (ASP) in children is 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 PRESENTTION

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 (orchidopexy) 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.

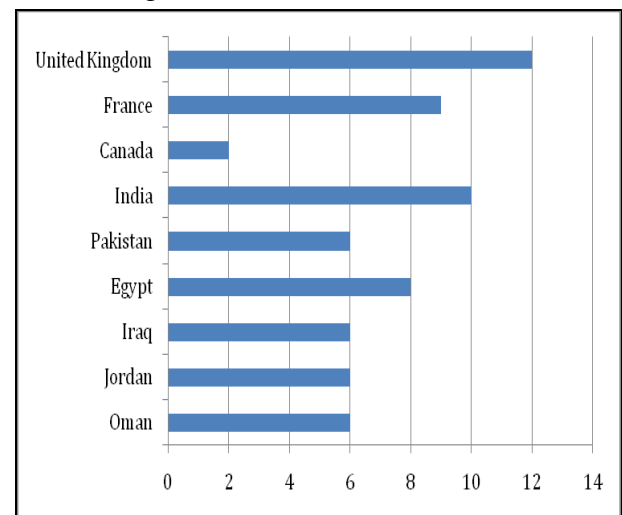


Figure 1: Distribution of practicing urologist in different countries

RESULTS

The response rate was 62% with 65 completed questionnaire returned (Figure2).

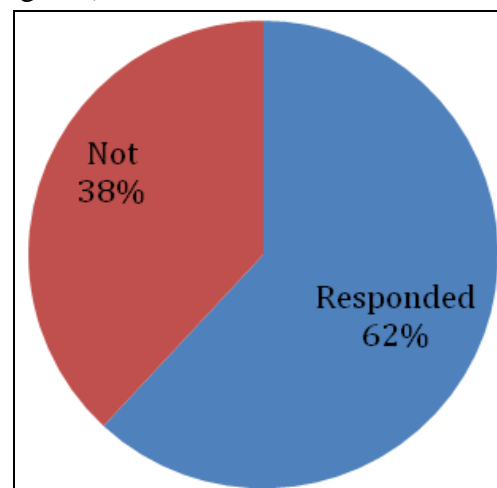


Figure 2: Survey Response

Of these, 22 (33%) were qualified consultants, while the rest were urologists in training (Figure 3).

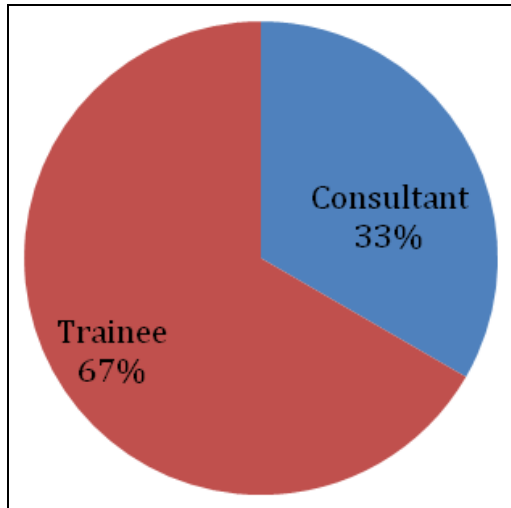


Figure 3: Urologists Level

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

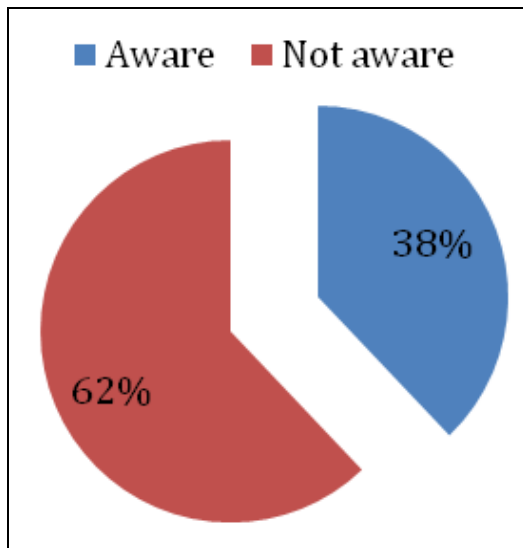


Figure 4: Awareness

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.

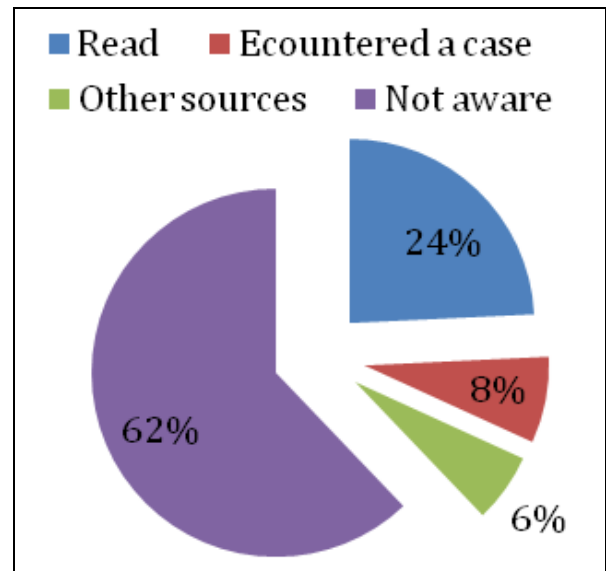


Figure 5: Source of Awareness

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).

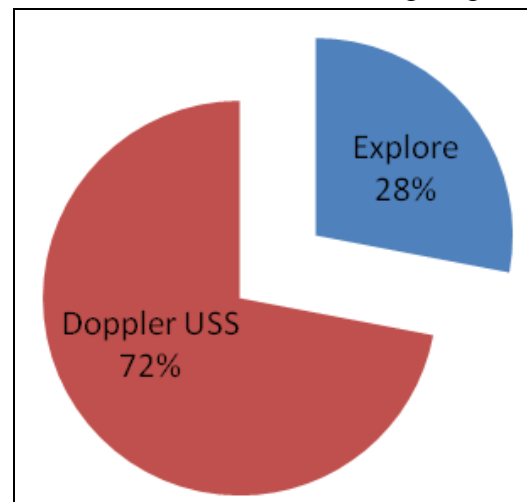


Figure 5: In those Aware

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.

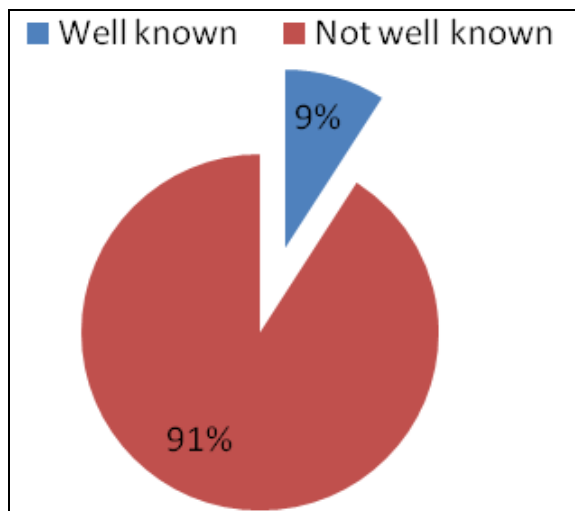


Figure 7: HSP-ASPS

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 children¹, 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 age². 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 Society³. 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 missed⁴. 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 1960⁵. Scrotal involvement in HSP mimicking testicular torsion is a self-limiting, benign disease and, if required, will respond to corticosteroid and/or antibiotic therapy⁴.

An acute scrotum is defined as an acute painful swelling of the scrotum or its contents, which is accompanied by local signs or general symptoms⁶. 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 grounds⁷. Thus, most patients undergo exploration⁸. 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 diseases⁹. 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 exploration¹⁰. A note of caution is appropriate, as three cases of torsion of a testicle or its appendix have been identified in patients with HSP^{11, 12}.

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.

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ثوختة

نەطەرەکی دەطمەن یی نیشانا هیلکا یا لئضر ل دەف زاروکا، ئیشکیشکرنا جورى نەخوشیی، دیارکرنا ئیزانینا ذ جھین باووری ئیکری

ئیشەکی: نیشانا دەطمەن یا هیلکا لئضر ئیک ذ نەخوشیین تەنطازن بو نشتەرطەتیا جوبارین میزی، کو دظیتن بهیئە دیارکرنا ل دەف زاروکا بەلکی هینوش شونلن بوربورا (هشب) بهیئە نیاسین ئیک ذ دیاردین نیشانا دەطمەن یا هیلکا لئضر، ئەوا بەلکی بهیئە نیاسین خاربونا هیلکی. ئەط شلوژەکرنا هاتە ئیشکیش کرن بو دوو جورال کارئ مە یی تزیشکی، دطەل دیارکرنا بنەجھین ئیزانینا کو دطریداینە ب نیشانا دەطمەن یا هیلکا لئضر و هینوش شونلن بوربورا ل لایى نوذدارین جوباریت میزی.

ریکین ظەکولینى: 9 ترسیار هاتن فریکرن بو 105 نوذدارین جوباریت میزی ئەوین هاتینە هەلبزارتن بشیوئەکی ل 9 وەلاتان کار دکەن و دطەلدا وەلاتی بریتانیا بو زانین ئەف نوذدارە هات بونە ظان نەذدارا هەمیایا کار کری ل دەژطەهین باووری ئیکری بو راهیئانی دضارضوئی طی شلوژەکرنا دا.

ئەنجام: ریژەتیا وەلامان دانا ترسیارا 62% مە 65 وەلامین تەمام وەرطرتن ذ نوذدارین جوباریت میزی. هەر وەسان دیار بوو کو 25 (38%) ذ نوذدارا ئیزانین رابردوو هەبوون ل سەر ئەیوئەندیا ناظ بەرا هینوش شونلن بوربورا و نیشا هیلکا لئضر یا دەطمەن، و ناظ بەرا کەسین ئیزانین هەین دور طی ئەیوئەندیی. هاتە دیارکرنا کو (28%) ذ نوذدارا رازیبون ب دیارکرنا بلز، بەلای هەندەک نوذدار ذیک داخاز کر کو دوپلر بو بهیئە کرن بەری ض بریار بهیئە دان ل دوماهیی 91% ذ نوذدارا یین دطی بروطرامی کار کری دیارکەن کو وان ض هەست ب ئەیوئەندیا هەردوو جورا نەدیت هەر وەسان 82% ذ وان نوذدارا دیارکر کو دظیت نوذدارى جوباریت میزی ئیزانین هەین ل دور طی دیاردی.

دەرتەنجام: ل سەر ذیک جدابونا ریکین ضارەسەتیا نیشانا هیلکا لئضر بەلای ئەیوئەندیا طی نیشی دطەل هینوش شونلن بوربورا نەیا نیش ضاف بوو طەلەک. ذبەر کو ضارەسەتیا نەخوشین نیشا هیلکا لئضر دیارکرنا بلەزە یا طونی بەلای دظیت نوذدارین جوباریت میزی ئیزانین هەین ل سەر هەر دوو جورا.

الخلاصة

سبب نادر لآلم الخصية الحاد لدى الاطفال؛ تقديم حالة مرضية، تقييم استبيان واستعراض المراجع

الخلفية والأهداف: ألم الصفن الحاد في الأطفال هو حالة طارئة في جراحة المسالك البولية، والتي عادة ما تستدعي إستكشاف الصفن. في الأطفال يمكن أن يظهر هينوش شونلن بوربورا (هشب) في بعض الأحيان كحالة ألم الصفن الحاد مع وذمة، والتي قد تشخص خطأ على أنها التواء الخصية. قدمنا في هذا البحث حالتين واجهتنا في ممارستنا الطبية؛ مع استعراض المراجع وتقييم الوعي بالصلة بين ألم الصفن الحاد و هينوش شونلن بوربورا لدى أطباء المسالك البولية.

طرق البحث: تم إرسال استبيان مكون من ٩ أسئلة إلى ١٠٥ من أطباء المسالك البولية الذين تم إختيارهم بشكل عشوائي والذين يعملون في تسعة بلدان مختلفة من بينها المملكة المتحدة علما أن جميع هؤلاء الأطباء عملوا في مؤسسات معتمدة للتدريب اثناء إجراء الإستبيان.

النتائج: كان معدل الاستجابة للإستبيان ٦٢٪ حيث تم الحصول على ٦٥ إجابة كاملة للإستبيان من أطباء المسالك البولية أعلاه. تبين أن ٢٥ (٣٨٪) طبيباً كانوا على علم مسبق للعلاقة بين هينوش شونلن بوربورا و ألم الصفن الحاد ، وبين أولئك الذين كانوا على بينة بهذا الارتباط تبين أن (٢٨٪) منهم كانوا يفضلون الإستكشاف الصفني الفوري ، في حين أن باقي الأطباء فضلوا إجراء فحص الموجات فوق الصوتية (دوبلر) قبل اتخاذ القرار. عموماً تبين أن ٩١٪ من الأطباء الذين إستجابوا للإستبيان شعروا بأن العلاقة بين هاتين الحالتين لم تكن معروفة جيداً وأن ٨٢٪ من هؤلاء الأطباء يعتبرون أن هكذا حالة هي قضية هامة على طبيب المسالك البولية أن يكون على بينة منها.

الاستنتاجات: على الرغم من الاختلافات في الممارسة 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 الإستكشاف الصفني الفوري نعتقد أنه يتعين على أطباء المسالك البولية أن يكونوا على دراية وعلم بالصلة بين هاتين الحالتين.