Portal vein thrombosis – a primer for the general physician

Editor – I was surprised to find the authors of ‘Portal vein thrombosis – a primer for the general physician’ strongly advocated an extensive search for thrombophilic conditions.\(^1\) While these conditions undoubtedly increase the risk of portal vein thrombosis, the question is does knowledge of these mutations alter the subsequent management of the condition or the duration of treatment?

There may be a justification in testing for some thrombophilic conditions, eg identifying myeloproliferative disorders with the JAK2 mutation, and some thrombophilias convey a higher risk of recurrence than others,\(^2\) but there is certainly little justification in screening for factor V Leiden and prothrombin gene mutations.

The 2012 National Institute for Health and Care Excellence guidelines for venous thromboembolism (VTE), which are surprising supportive of thrombophilia screening, do not include factor V Leiden and the prothrombin mutation as they do not increase the risk of recurrence to a clinically significant extent.\(^3\)

In contrast, the 2010 British Society of Haematology guidelines state ‘testing for heritable thrombophilia after a first episode of intra-abdominal vein thrombosis has uncertain predictive value for recurrence. Grade C evidence – as no studies have investigated how the finding of a heritable thrombophilia should influence management’.\(^4\)

Analysis of the large multiple environmental and genetic assessment study showed that testing for inherited thrombophilia did not reduce recurrence of venous thrombosis.\(^5\)

The American College of Chest Physicians guidelines on VTE, which recommend ongoing anticoagulation after an unprovoked VTE, list thrombophilias among factors that predict risk of recurrence, ‘but not strongly enough to influence recommendations on duration of therapy’.\(^6\) And US guidelines have an equally clear message of ‘do not perform thrombophilia testing in patients following an episode of unprovoked VTE’.\(^2\)

In summary, thrombophilia is commonly evaluated in patients without a clear indication for testing and not only that, but frequently during times when the results may be unreliable.\(^7\)

Conflicts of interest

The author has no conflicts of interest to declare.

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References


An unusual case of orthopnea

Editor – I read with interest the case of diaphragmatic paralysis presented by Keelan et al.\(^1\) As they stated in their article, an iatrogenic cause should be considered; however, one they failed to mention, which is important for the physician to be aware of, is following pulmonary vein isolation for the treatment of atrial fibrillation.

The number of pulmonary vein isolations being performed is steadily increasing.\(^2\) At present, two main strategies exist: point by point ablation with radiofrequency energy or freezing using an expandable balloon catheter (cryoballoon). Although both are associated with phrenic nerve palsy, cryoballoon ablation has the higher complication rate reported over a number of studies (4.6–11.2% versus 0–0.3%).\(^3\) The majority of complications result in a temporary paralysis with an average recovery time of 4 months; however, permanent paralysis has been recognised.\(^4\) A right-sided unilateral palsy is the commonest reported because of the proximity of the right phrenic nerve to the right-sided pulmonary veins (especially the right superior vein). Intra-procedural phrenic nerve stimulation to monitor for complications during cryoballoon ablation has cut the rates of injury significantly\(^5\) and is routinely used at our centre.

Iatrogenic phrenic nerve palsy following pulmonary vein isolation can be easily overlooked as a potential cause both by the patient and clinician, particularly when the presentation is weeks after the procedure and our medical admissions units are frequented by breathless patients with exacerbations of chronic lung disease (personal experience). A higher index of suspicion should be employed, with earlier use of appropriate investigations.

Conflicts of interest

The author has no conflicts of interest to declare.

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References

Letters to the editor

An unusual case of orthopnea

Editor – Keelan et al describe an interesting patient with bilateral phrenic nerve palsies, which they ascribe to cervical spondylody.

The phrenic nerve arises mostly from C4 with contributions from C3 and C5. It would be quite exceptional, if not anatomically impossible, for cervical spondylody to affect only those fascicles destined for the phrenic nerves, without any clinical evidence of a myelopathy or other radicular signs. They cite eight reported cases, seven of which had a myelopathy and one that was unilateral. A much more likely diagnosis is neuralgic amyotrophy, which may be bilateral in up to 30% of patients and may follow strenuous exercise (17%), as in this patient who was lifting heavy iron tables the day before the onset of symptoms. Phrenic nerve palsy, both unilateral and bilateral is well described in neuralgic amyotrophy and maybe the presenting and only feature.

Conflicts of interest

The author has no conflicts of interest to declare.

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References


Adrenal insufficiency – recognition and management

The observation that Addison’s disease may present as an unexplained reduction in insulin requirement in an individual with diabetes mellitus has, as its corollary, the observation that a requirement for an unusually large dose of hydrocortisone (over and above concurrent fludrocortisone therapy) to maintain an adequate blood pressure (BP) might signify that Addison’s disease coexists with hitherto unrecognised thyrotoxicosis. This was the case in a 74-year-old man who had initially presented with Addisonian crisis characterised by BP 70/40 mmHg and a pulse rate 130 bpm. The diagnosis of Addison’s disease was subsequently validated by a positive synacthen test. Nevertheless, despite the fact that hydrocortisone was co-prescribed with fludrocortisone, he required as much as 80 mg/day of hydrocortisone to maintain a BP 120/80 mgHg. The fact that tachycardia also persisted raised the index of suspicion for thyrotoxicosis, a diagnosis that was duly validated by free thyroxine and tri-iodothyronine levels of 45.3 nmol/L (normal 10–30) and 5.9 nmol/L (normal 0.8–3), respectively. A flat response to the thyrotropin-releasing hormone test clinched the diagnosis. Following treatment with carbimazole he became euthyroid and his pulse rate fell to 68 bpm. It also subsequently became possible to reduce the dose of hydrocortisone to a level of 30 mg/day, which maintained him in good health.

A comparable scenario was documented in a 42-year-old woman in whom the initial diagnosis was Addison’s disease and in whom treatment with prednisolone 5 mg twice per day resulted in a 1.5-year period of relief of symptoms. Subsequently, however, she experienced two episodes of Addisonian crisis 4 months apart. The maintenance dose of prednisolone was then increased to 10 mg in the morning and 5 mg in the evening, and this was co-prescribed with fludrocortisone 50 µg/day. Following identification of thyrotoxicosis as the precipitating cause of adrenal crisis, she was rendered euthyroid by means of carbimazole, followed by radioiodine. After she became euthyroid, she remained symptom free and gained 8 kg in weight while taking prednisolone 5 mg/day and fludrocortisone 50 µg/day.

Although the association of Addison’s disease and thyrotoxicosis is rare,2–5 there should be a heightened index of suspicion for coexisting thyrotoxicosis when symptoms and signs of hypoadrenalism persist despite progressively increasing doses of replacement therapy or when a previously well managed patient experiences Addisonian crisis without an obvious precipitating cause.

Conflicts of interest

The author has no conflicts of interest to declare.

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References