Sigmoid Sinus Diverticulum, Dehiscence, and Venous Sinus Stenosis: Potential Causes of Pulsatile Tinnitus in Patients with Idiopathic Intracranial Hypertension?

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ABSTRACT

BACKGROUND AND PURPOSE: Pulsatile tinnitus is experienced by most patients with idiopathic intracranial hypertension. The pathophysiology remains uncertain; however, transverse sinus stenosis and sigmoid sinus diverticulum/dehiscence have been proposed as potential etiologies. We aimed to determine whether the prevalence of transverse sinus stenosis and sigmoid sinus diverticulum/dehiscence was increased in patients with idiopathic intracranial hypertension and pulsatile tinnitus relative to those without pulsatile tinnitus and a control group.

MATERIALS AND METHODS: CT vascular studies of patients with idiopathic intracranial hypertension with pulsatile tinnitus (n = 42), without pulsatile tinnitus (n = 37), and controls (n = 75) were independently reviewed for the presence of severe transverse sinus stenosis and sigmoid sinus diverticulum/dehiscence according to published criteria. The prevalence of transverse sinus stenosis and sigmoid sinus diverticulum/dehiscence in patients with idiopathic intracranial hypertension with pulsatile tinnitus was compared with that in the nonpulsatile tinnitus idiopathic intracranial hypertension group and the control group. Further comparisons included differing degrees of transverse sinus stenosis (50% and 75%), laterality of transverse sinus stenosis/sigmoid sinus diverticulum/dehiscence, and ipsilateral transverse sinus stenosis combined with sigmoid sinus diverticulum/dehiscence.

RESULTS: Severe bilateral transverse sinus stenoses were more frequent in patients with idiopathic intracranial hypertension than in controls (P < .001), but there was no significant association between transverse sinus stenosis and pulsatile tinnitus within the idiopathic intracranial hypertension group. Sigmoid sinus dehiscence (right- or left-sided) was also more common in patients with idiopathic intracranial hypertension compared with controls (P = .01), but there was no significant association with pulsatile tinnitus within the idiopathic intracranial hypertension group.

CONCLUSIONS: While our data corroborate previous studies demonstrating increased prevalence of sigmoid sinus diverticulum/dehiscence and transverse sinus stenosis in idiopathic intracranial hypertension, we did not establish an increased prevalence in patients with idiopathic intracranial hypertension with pulsatile tinnitus compared with those without. It is therefore unlikely that these entities represent a direct structural correlate of pulsatile tinnitus in patients with idiopathic intracranial hypertension.

ABBREVIATIONS: IIH = idiopathic intracranial hypertension; PT = pulsatile tinnitus; SSDD = sigmoid sinus diverticulum/dehiscence; TSS = transverse sinus stenosis

Pulsatile tinnitus (PT) is the perception of an abnormal sound that is synchronous with the heartbeat. The phenomenon may result from abnormal mechanical somatosound production (due to abnormal turbulence or location of vascular flow) or enhanced perception (due to increased transmission of the somatosound or reduced conduction of normal sound).1 PT is a well-recognized feature of idiopathic intracranial hypertension (IIH), being present in approximately 60% of patients.2,4 The pathophysiology of PT in these patients is unknown; however, observed variations in transverse and sigmoid sinus anatomy seen in IIH have been proposed as a possible underlying cause.

Transverse sinus stenosis (TSS) is well-described as a cause of PT, and resolution of symptoms has been reported following successful venous sinus stent placement.5-8 Venous sinus stent placement performed as a treatment for IIH has also resulted in resolution of PT in affected patients,7,8 which lends support to the
therefore essential to enable appropriate targeting of therapy.

IIH.

to patients with IIH without PT and a control group without
and SSDD was increased in patients with IIH with PT relative
for evaluation. When
Patients were excluded if there was no CT venogram available
of IIH,1,14,15 leading some authors to suggest that SSDD may re-
the modified Dandy criteria20 in all except 1 patient in whom
flect a manifestation of subclinical IIH.2,14
Interventional studies have revealed that endovascular
stening of TSS or surgical correction of SSDD may resolve
brane responsible for PT in IIH.9,10 However, PT is not seen
in all patients with IIH with TSS, and other factors may be re-
quired for the symptom to manifest.

More recently, sigmoid sinus diverticulum and/or dehiscence
(SSDD) has been shown to be associated with PT.1,11-16 This
association has been noted to be particularly frequent in patients
with elevated body mass indices and those with imaging features
of IIH,1,14,15 leading some authors to suggest that SSDD may re-
reflect a manifestation of subclinical IIH.2,14

We aimed to determine whether the prevalence of TSS
and SSDD was increased in patients with IIH with PT relative
to patients with IIH without PT and a control group without
IIH.

MATERIALS AND METHODS

Subjects

One hundred seventy consecutive patients diagnosed with IIH
were identified from a clinical data base at our institution and
selected for retrospective review. IIH diagnosis was made with
the support of CSF pressure measurements and according to
the modified Dandy criteria20 in all except 1 patient in whom
lumbar puncture was precluded due to cerebellar tonsillar herni-
ation. Case notes were reviewed for demographic and clinical
details, including a history of pulsatile tinnitus. Laterality of
PT was not consistently recorded and hence not documented.
Patients were excluded if there was no CT venogram available
for evaluation. When >1 CT venogram had been obtained on
the same patient, the examination closest to the diagnosis of
IIH was selected for analysis. Seventy-nine patients with IIH
(mean, 32 ± 5.8 years of age; male/female ratio, 4:75) were
available for evaluation.

A cohort of control subjects was generated by using contemporaneous CT
angiograms obtained on the same CT scanner for reasons other than suspected
venous pathology or pulsatile tinnitus. A list of CTA examinations was generated
by searching the PACS archive for intracranial CTAs performed in the study pe-
riod (January 1, 2005, to May 22, 2015) that contained the text “normal” in the
body of the report. The resulting 1250 studies were ranked by age with subjects
younger than 16 and older than 45 years of age excluded to closely model the IIH
cohort. The report text of the remaining scans was manually screened, and stud-
ies with abnormal findings were excluded. The resulting 183 CTAs were re-
viewed, and further exclusions were made if there was confounding pathol-
ogy or if venous opacification was insufficient (defined as <160 HU measured at the torcula). After this
process, 75 control subjects were available for evaluation.

The study was reviewed by the local National Health Service
Research Ethics Committee and was considered to represent “ser-
vice evaluation.”

Radiologic Assessment

CTAs and CT venograms were obtained with a LightSpeed 16
scanner (GE Healthcare, Milwaukee, Wisconsin). Scanning pa-
rameters were 0.625-mm collimation; pitch, 0.938:1; noise index,
2.2; automaticmepicameter limit, 380.

The presence of venous sinus stenosis, sigmoid plate dehis-
cence, and diverticula was assessed by 2 neuroradiologists inde-
pendently (J.A.L. and M.R.E., with 9 and 15 years of radiology
experience, respectively). Venous sinus luminal dimensions were
first evaluated with maximum-intensity-projection reconstruc-
tions to provide an overview of the narrowest portion of the sinus
encountered from the torcula to the distal sigmoid sinus. Multi-
planar reformats (0.6 mm thick) at standard magnifications and
perpendicular to the plane of the skull vault were then used to
assess the height of the sinuses. Percentage stenoses were calcu-
lated as the ratio of the maximal narrowing relative to the maxi-
mal height of the superior sagittal sinus (measured 2 cm above the
torcula, Fig 1). Stenoses were categorized as >50% and >75%
relative to the superior sagittal sinus according to previously pub-
lished criteria.21

A rigorous, objective radiographic classification of SSDDs is
yet to be established. For this study, we adapted a previously de-
scribed methodology,1 using MPR reconstructions and an edge-
sharpening tool available on our PACS. Assessments were made
with standard bone windows (width, 4000 HU centered at 400
HU). Dehiscence was characterized as complete absence of corti-
cal bone adjacent to the sigmoid sinus (air-on-sinus sign) evident
on at least 2 consecutive 0.6-mm sections in 2 orthogonal planes
(Fig 2). “Sigmoid sinus diverticulum” was defined as a focal out-
pouching of the normal semicircular sigmoid sinus groove ex-

FIG 1. A, Superior sagittal sinus height measurement (2 cm above the torcula). B, Transverse sinus
stenosis measurement.
Consensus review was undertaken for all discrepant cases derived from the initial independent readings.

**Statistical Analysis**

Data were entered into an Excel spreadsheet (Excel for Mac, 2011; Microsoft, Redmond, Washington). The statistical package StatPlus (AnalystSoft, Walnut, California) was used for descriptive statistics.

The prevalence of TSS (>75% and >50%), sigmoid sinus dehiscence, and sigmoid sinus diverticulum in patients with IIH was compared with that in the control group to see whether the previously reported associations were replicated in our cohort. Further subgroup analyses were performed to compare the PT-IIH group with the controls and the non-PT IIH group. Group comparisons were also performed to compare the prevalence of ipsilateral TSS and SSDD in the cohorts in case simultaneous abnormalities were needed for PT to manifest. Statistical comparison was performed with χ² or Fisher exact tests as appropriate, with P < .05 being required for significance.

We performed further exploratory between-group comparisons, taking into account variations in the laterality of TSS and SSDD (ie, left, right, unilateral, bilateral, and either unilateral or bilateral).

**RESULTS**

There was no significant difference in the average age of the IIH group (32 ± 5.8 years) and control group (32 ± 10.2 years). Of the 79 patients with IIH, 42 had PT. The average body mass index in the IIH group was 37, and there was no significant difference in body mass indices between patients with IIH with and without PT. The IIH group was almost exclusively female (75:4), whereas the control group comprised 35 men and 40 women.

The characteristics and imaging analysis results of the groups are summarized in the Table.

Patients with IIH were significantly more likely to have a severe grade of venous sinus stenosis (>50% and >75%) compared with controls. When both left and right transverse sigmoid sinus drainage pathways were considered together, 49% of patients with IIH had >75% stenosis affecting 1 side compared with just 7% of controls (P < .001). This significance was maintained for the PT-IIH subgroup analysis, but there was no significant association between severe dural venous sinus stenosis and PT within the IIH group.

Dehiscence, as defined by the "air-on-sinus" sign, was more common on the right in all groups. In the IIH group, 41% of all temporal bones were dehiscent, compared with 27% of controls (P = .01). The significance of this difference was retained when controls were compared with the PT-IIH subgroup, but there was no significant difference between patients with and without PT in the IIH group.

Sigmoid sinus diverticula were twice as common in patients with IIH compared with controls (8% versus 4%); however, the difference was not significant, and no other significant differences were found in the subgroup analyses.

There was no significant difference in the prevalence of ipsi-
Patients with PT and SSDD have been found to have clinical and imaging characteristics of IIH. A number of mechanisms have been proposed to explain this association. It is possible that the impact of pulsatile, turbulent blood flow on the sinus wall could cause the formation of SSDD. Alternatively the presence of SSDD may be a direct result of raised CSF pressures. It has also been postulated that sigmoid sinus dehiscence could cause PT in these patients due to increased transmission of turbulent flow to the inner ear after loss of the insulating properties afforded by cortical bone. Although our findings did demonstrate an association between dehiscence and IIH, there was no association within the IIH group to support a direct causal relationship. We also investigated the suggestion that a combination of TSS and SSDD may be required to cause PT, but we found no increased frequency of ipsilateral TSS and SSDD in our PT cohort with IIH to support this.

It is possible that dehiscence involving specific locations on the curve of the sigmoid sinus results in PT symptoms. We did not distinguish different types of stenosis: tapered “extraluminal” narrowing versus “intraluminal” narrowing typified by arachnoid granulations. This could also have limited our ability to detect a significant association between stenosis and PT.

It is unclear whether sigmoid sinus dehiscence and dehiscence represent separate entities or whether there is an underlying pathophysiologic spectrum of disease. Diverticula may cause turbulent venous flow and lead to symptomatic PT when coexisting with elevated intracranial pressure, independent of TSS; how...
ever, our IIH cohort did not reveal any association between PT and diverticula to lend support to this theory.

We explored the combined effect of TSS and ipsilateral SSDD, but it is possible that a more complex combination of anatomic features is required to cause PT or that the pathophysiology differs between patients. It has been shown that ipsilateral venous outflow dominance, high jugular bulb, and temporal bone hyperpneumatization may also be important in the manifestation of SSDD as PT.13,16 Complex interactions with venous dominance, the site and size of dehiscence, and the type of stenosis (intraluminal versus extraluminal) were not fully evaluated in our study. Although we did not document venous dominance directly, diverticula and dehiscence were observed more frequently on the right; this feature is of interest because this is more likely to be the dominant sinus.31

Our findings reveal a high prevalence of PT in IIH, which is in line with previous studies. The data also corroborate previous research indicating a high frequency of transverse sinus stenosis and sigmoid sinus wall abnormalities in IIH. However, we found no significant association between PT and venous sinus or sinus wall abnormalities within the IIH group, suggesting that a direct causal link is unlikely. Furthermore, ipsilateral dehiscence and TSS abnormalities were not significantly more frequent in the PT group; this finding suggests that even when combined, these factors are not sufficient to cause PT.

Our dataset contained a large validated cohort of patients diagnosed with IIH, corroborated by CSF pressure measurements; however, the study design has some important limitations. First, the retrospective nature meant that there was nonuniformity of data. Most important, we had limited information on the laterality of PT. It is recognized that PT in IIH is most commonly unilateral, and this information would have allowed a greater confidence in relating PT to the venous correlates. The lack of well-defined criteria and the subjective nature of the scoring for SSDD are also potential drawbacks; however, these were similarly applied across groups with standardized viewing parameters, and a consensus review was undertaken in cases of disagreement.

Another potential bias was introduced by including only patients with IIH who had undergone CT venography, which a large number of patients in the data base had not undertaken. The use of CTAs for the control group meant that observer blinding was not possible, and cases could be distinguished from controls. Nevertheless, observers were blinded to the presence of PT within the IIH group, and the question of this association was the primary aim of this study.

No previous studies describe the incidence of SSDD/TSS in confirmed IIH cases, to our knowledge; therefore, we were unable to perform a power calculation, and the sample size was determined by the size of the clinical data base.

Finally, the control group was not matched for sex, resulting in a female bias for the IIH cohort. Previous control cohorts have comprised subjects with middle ear symptoms referred for ear, nose, and throat investigations. The relatively high proportion of SSDD abnormalities found in these subjects (up to 18%) may misrepresent the general population. We thought it was important to know the prevalence of SSDD in the general population to better understand the likelihood of SSDD as a coincidental finding. A mixed-sex control group was therefore used to provide data about the incidence of SSDD.

Four percent of controls in our cohort had a combined conduit score of <5, in line with the 7% of controls in the study by Grewal et al.13 Sigmoid sinus dehiscence was more common in our control cohort (44% of patients, 53% of ears) compared with 1.2% and 18% of incidental SSDD reported in previous studies.2,14 This discrepancy is likely to reflect shortcomings of CT assessment. Only tiny focal areas of apparent bone loss were required to be categorized as dehiscence; however, as previously shown in temporal bone histologic studies, bone thickness of <0.1 mm can be falsely read as dehiscent.33,34 We also found a higher incidence of sigmoid sinus diverticula in our control cohort than described in previous studies: 8% compared with 0.5% reported by Grewal et al.,14 which may reflect differences in the demographic studied or possibly be due to sampling bias inherent in our smaller sample size.

CONCLUSIONS

Although TSS and sigmoid sinus dehiscence were shown to be more frequently present in patients with IIH compared with controls, our findings do not support the proposed pathophysiological mechanism that SSDD or TSS are directly responsible for PT in IIH. The finding of SSDD in a patient with PT may be coincidental, and patients considering surgical intervention should be counseled accordingly.


REFERENCES


23. Shulman A. Brain pulsatility and pulsatile tinnitus: clinical types. *Int Tinnitus J* 2012;17:2–3 Medline