



Spina Bifida Occulta in a Nigerian Tertiary -Care Hospital

Uduma Felix U^{1*} and Okoye IJ²

Abstract

Background: Spina bifida occulta (SBO) is a congenital midline defect of fusion of the posterior vertebral arch. It has no associated protrusion of spinal cord or meninges.

Purpose: To analyze a single-institution study on spina bifida occulta.

Methods: The studied period was from 27th June, 2012 to 31st October, 2013. Consecutive Patients who came to Radiology Department, University of Uyo Teaching Hospital, Uyo, Nigeria for cervical spine radiographs were prospectively studied. For each patient, anterior-posterior (AP) and lateral cervical spine radiographs were taken according to standard protocol (5 cm above the suprasternal notch and 2.5 cm posterior to mandibular angle for AP and lateral centring points respectively). Results were analyzed using SPSS 13.0 for Windows software package.

Results: One hundred and forty patients (age up to 89 years) were studied with 82 males and 58 females. Eighteen patients had SBO giving an incidence of 12.86%. Males were 10 and females were 8 giving a M: F ratio of 1.25:1. However, the % age of males with SBO with respect to total studied male population was 12.19% but 13.79% among females. All positive cases of SBO were central in location and the size of all neural arch defects were less than 1cm wide. The commonest pattern of SBO was solitary with 66.67%. 100% of females had solitary SBO, localized at C7 and devoid of any attending spondylosis.

In general, 26 SBO were seen in 18 patients due to multiplicity of SBO in a patient. Patients with SBO who also had associated spondylosis were 22.22% and this was only peculiar to males.

Conclusion: Incidence of cervical spina bifida occulta in University of Uyo teaching hospital is 12.86%. It has a female bias, lower cervical vertebra preference and central localization restriction. Multiplicity of defects tends to be associated with spondylosis.

Keywords

Spina bifida occulta; Cervical defects; Vertebral arch; Spondylosis

Introduction

Spina bifida literally connotes a cleft in the spinous process of a vertebra. In essence, it is a “spine in two parts” or “open spine” or “bifid spine” whether there is a protrusion of the spinal structures or not [1-3]. Spina bifida is the commonest subtype of the wide spectrum of neural tube defects (NTD) [1,4]. NTD on its own is a spinal dysraphism which is a congenital anomaly of the spinal column [5].

It involves the incomplete development of the brain and/or spine [4].

The history of spina bifida ran into centuries. The foremost description of spina bifida dated back to 1587 by Peter Van Forest who subsequently carried out the first reported surgical procedure of meningocele sac ligation in 1610 [6]. However, Nicholas Tult noted spina bifida occulta as a congenital developmental defect in 1642 [7]. Subsequently the term ‘spina bifida occulta’ was coined by Virchow in 1875 on his evaluation of a case of spina bifida concealed under the skin [2]. This brings to mind, the broad classification of spina bifida into spina bifida occulta (SBO) and spina bifida aperta (spina bifida cystica) [1,6]. The distinction is not farfetched. As the later has a visible protrusion of spinal cord and/or meninges via a defect in the spine [1,6,8]. However our scope is limited to spina bifida occulta.

SBO is a congenital midline defect of fusion of the posterior vertebral arch without protrusion of the spinal cord or meninges [1,5,8-10]. It is a milder and commoner condition, characterized by non-fused spinous processes and lamina with an intact dural sac. Essentially, it has normal skin cover [1]. The cutaneous and subcutaneous evidence of SBO are hypertrichosis, palpable bony defect, dimples, hyper pigmentation, scarring, haemangioma, telangiectasis, lipoma and linear scleroderma [11].

SBO occurs more often in the lumbosacral junction compared with the cervical spine [10].

SBO arises from failure of the neural tube to close during the fourth week of embryogenesis with consequent failure of the bones around the spinal cord to close [4,8]. Sonographic studies have shown 3 spinal ossification centres (vertebral body and each lamina-pedicle junction) [2]. The developmental stages of the vertebra are precartilaginous, chondrification and ossification [12]. Precartilaginous stage involves migration of sclerotome cells ventromedially (to form vertebral body), dorsally (neural arch) and ventrolaterally (costal process) [12]. Posterior arch chondrification starts at the pedicle at 6th intrauterine week and ends at the midline at 4th month [12]. This is followed by cartilaginous arch ossification at 3-4th year of life [12]. Failure of chondrification or ossifications results in posterior arch defects [12]. Chromosomal abnormalities and single gene disorders have been implicated in the aetiology of SBO [6]. Many malformations have been associated with spina bifida and this arose from damage to mesodermal or endodermal anlagen caused by over-distention of the embryonic neural tube [6]. Syndromic associations of SBO are Larsen’s syndrome, Atelosteogenesis Type III, Arnold Chari’s malformation, Gorlin’s syndrome, Sprengel shoulder, neurenteric cysts [13-18]. Larsen’s syndrome is a rare genetic disorder with ligamentous hyperlaxity, abnormal facial features, multiple joint dislocations and long cylindrical fingers that do not taper [14,19]. SBO of the cervical or thoracic vertebrae is found in 45-60% of cases in Gorlin’s syndrome, a dominantly inherited condition [15].

Generally, spina bifida occurs more frequently in females and more often in Hispanic populations [4]. Risk factors of SBO include a previous pregnancy with fetal NTD [4]. Others are insulin-dependent diabetes, anti-seizure medications, obesity and high temperatures in early pregnancy [4].

*Corresponding author: Felix U Uduma, Department of Radiology, Faculty of Clinical Sciences, University of Uyo, Nigeria, E-mail: felixuduma@yahoo.com

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Objective

The objective of this study is to analyze a single-institution contemporary experience with spina bifida occulta.

Materials and Methods

The study took place between 27th June, 2012 and 31st October, 2013. Consecutive patients who came to Radiology Department, University of Uyo Teaching Hospital, Uyo, Akwa Ibom State, Nigeria for cervical spine radiographs on any account were prospectively studied. Their clinical and demographic data were extracted; patients' consents were sought prior to radiographic examinations. For each patient, anterior-posterior (AP) and lateral cervical spine radiographs were taken according to standard protocol.

In AP position, the patient lies supine in the centre of X-ray couch with the back of the head rested on the couch top. It is ensured that the head is not rotated through immobilization using sandbags. Then the chin is elevated to the point that the lower border of the mandible is 90° to the detail screen cassette. Marker is applied, beam collimated and protection also applied. Centring point is 5cm above the suprasternal notch and in the midline. The central ray is directed vertically with the tube angled 15° cephalad. Exposure is done under arrested respiration.

In the lateral position, patient stands or sits in the true lateral position, against the erect cassette holder. The chin is raised such that the angle of the mandible is clear of cervical vertebrae. The shoulder is depressed as low as possible. The tip of the shoulder is rested on the lower border of cassette. Head is immobilized, marker applied, beam collimated and protection applied. Centring point is 2.5cm posterior to the angle of the mandible. The central ray is directed horizontally at 90° to the cassette. Exposure is done under arrested respiration the 7th cervical vertebra is shown with the arm in weight bearing. Focal film distance applied is 150cm to make up for the large object- film distance.

In some cases, anterior-posterior open mouth view was also done to properly show cervical vertebrae 1-3.

Any identifiable cervical posterior arch defect was evaluated with respect to vertebral localization, central defect or not, size of defect measured with metric ruler. Features of spondylosis like disc space narrowing, osteophytosis, endplate sclerosis and vacuum phenomenon were sought for.

Exclusion criteria included non-optimal radiographs, non-standardized radiographs and patients without complete bio-data. Results were analyzed using SPSS 13.0 for Windows software package (SPSS, Chicago, Ill).

Results

A total of 140 patients were studied with 82 males and 58 females giving a male to female ratio of 1.4:1. Age range was between 10 and 89years of life. The largest male population was 15.71% (n=22) in the 30-39 age range while the largest female population was 17.14% (n=24) in the 50-59 age range.

Eighteen patients had spina bifida occulta giving an incidence of 12.86%. Males were 10 and females were 8 giving a M: F ratio of 1.25:1. However, the percentage of males with SBO with respect to total studied male population was 12.19% whereas it is 13.79% among females. The youngest age in males and females with SBO on

presentation were 32 years and 40 years respectively. While the oldest age in males and females were 59 years and 69 years respectively.

In general, every defect in all positive cases of SBO seen in this study was central in location. The size of all neural arch defects were all less than 1cm wide.

The commonest pattern of SBO was solitary with 66.67% of positive cases and M:F ratio of 1:2. All (n=8,100%) of females participants had solitary SBO, 100% of this solitary SBO in females were localized at C7. All these females with SBO were devoid of any attending spondylosis. Forty% of males with solitary SBO were shared equally into C5 and C7 locations.

In general, 26 SBO were seen in 18 patients due to multiplicity of SBO in a patient.

The percentage of patients with positive SBO and associated spondylosis were 22.22 (n=4) and this is only peculiar to males. The components of spondylosis seen were only anterior osteophytes and disc space narrowing's.

Discussion

It is interesting to note that spina bifida occulta was not observed on the first, second and third cervical vertebrae in this study. This is due in part to the fact that open mouth view of the cervical spine was not routinely done in this study. As they were rarely included in the clinician request except in some cases of trauma. Recall that this view is specifically aimed at evaluating C1, C2 and C3 vertebrae [20,21]. Another reason for absent SBO in upper cervical vertebrae could be the rarity of this condition in the upper cervical vertebrae. A study discovered an incidence of high cervical lesions in only about 3.9% of cases [1]. Also rachischisis or SBO of the atlas is well recognized but rarely encountered [17]. For example, 30 skulls were evaluated for SBO of atlas in a study [22] and only 2 cases were detected. This low incidence of SBO in the upper cervical vertebrae could also not be unconnected with the general low incidence of SBO in cervical spine. The trend of defects in spinal dysraphism to which SBO is an integral component has cervical spine as the least in the order of frequency of occurrence [1]. In general population, SBO is more likely to occur at L5 or S1 level [5,9]. In India, lumbosacral region followed by lumbar region were the most common sites of spina bifida [6].

Spina bifida is known to be one of the most frequently occurring birth defects [11]. As such, one would expect it to be commonly seen in paediatric radiographs. But the earliest age of presentation for both sexes in this study was well into adulthood, 32years and 40years for males and females respectively. This is a study in which we have little or no contribution to patient recruitment as they were purely referrals to our department. Their late presentation could also be due to asymptomatic nature of SBO. SBO is the least severe form of spina bifida and in many patients, it causes no problems at all [4,10]. In some cases, it causes only minor physical disabilities, kyphosis or scoliosis [1,4,23]. They are actually in most cases seen as incidental radiographic findings especially when investigating the cervical spine following trauma [17]. Trauma is common but not the exclusive preserve of active, mobile middle aged people who are more prone to vehicular accident [24]. Cervical spine radiographs are also requested on account of neck or upper limb pains. These are mostly the corollary of age-related degenerative disc diseases. Thus by omission or commission we have excluded the paediatric age group from this study thereby denying us the opportunity of truly knowing the earliest age of radiographic detection of SBO.

In agreement with other studies, females who had SBO were more than males when computed with respect to each sex population [4]. In this study, the number of separate SBO exceeds the number of patients with SBO due to the SBO multiplicity in a single patient. Spina bifida occulta can involve one or several adjacent vertebral segments. Most commonly, the transitional levels of the spine [7]. Multiple SBO was only seen in males and seen in 60% of positive cases. This was shared into 40% of patients with 2 locations and 20% in 3 locations.

Cervical spondylosis was seen coexisting with SBO in some men. This was only observed in men with multiple SBO and 40% of men with SBO. Very high coincidence of SBO has been reported in more than 60% of lumbar spondylolysis, precedence to spondylosis [5]. The SBO may change the spine biomechanics thereby increasing the stresses at the pars interarticularis in spondylolysis [5]. This will culminate in spondylolisthesis and spondylosis.

The hospital based incidence of SBO in our study is 12.86%. This is somewhat close to 17% SBO occurrence rate seen on spine examinations in a study [11]. However the variation is wider when compared to another study with SBO incidence of 20% [5]. But this is comprehensible as the later was a general population based study. The reported frequency of occurrence of SBO varies widely, depending largely on the age groups included in a particular study [11]. It is estimated that approximately 70,000 people in the United States have Spina Bifida [4].

The imaging work up of developmental lesions of axial skeleton like SBO commences with conventional radiography [25]. This, we exploited in this study as it is affordable and available with little radiation exposure to the patients. Alternative modality would have been computed tomography (CT). It has good osseous anatomical definition, multi-planar reconstructive potentials and capacity to separate trauma insinuating as SBO. But, that would have added to patients' cost and increase radiation dose. Magnetic resonance imaging (MRI) assists in evaluations of intrinsic spinal cord parenchymal changes and the potential neural compression [25,26]. But its ability in assessing SBO a cortical bony lesion is not optimal. With some lesions, bony scintigraphy or PET scanning may be helpful to assess for metabolic activity [25]. With the prenatal screening, the incidence of apart is gradually declining, whereas the detection of occulta has increased with the advent of MRI [1].

Spina bifida occulta mat be considered to be of no clinical significance as it does not predispose the patient to spinal column fractures or cord injuries [10]. Therefore, it does not require any treatment except when there is an accompanying cervical spondylosis as seen in some of our patients. However, better maternal nutrition and pre-conceptional folic acid supplementation can be prophylactic. Research [4] has shown that ingestion of 400 micrograms of folic acid (especially before pregnancy) by a woman cuts her risk of neural tube defects by as much as 70%. Similarly, improved antenatal care and high-resolution ultrasound for prenatal screening and biochemical markers have also helped in reduction of spinal dysraphism [1].

The differential diagnoses of SBO are cervical spondylolysis, retro somatic cleft, bony destruction, C1 burst fracture of Jefferson, Burner "stinger" syndrome and atlas assimilation [3,10,17,27].

The significance of our study is to reawaken the consciousness of Radiologists, Neurosurgeons, Neurologists, Orthopedic surgeons and others to spina bifida occulta. SBO is frequently considered as an

incidental finding, and oft times not even mentioned in radiological reports [7]. SBO is also a less likely recognizable defect [6]. This may be due to lack of dedicated search on the part of the reporting Radiologist. We have also attempted to establish the incidence in our local environ through this study. The exact incidence of this congenital malformation is unknown because it is for the most part asymptomatic [10]. Being conscious of SBO will also necessitates the Radiologist searching for known syndromic associations like Larsen syndrome or attendant features like spondylosis no matter how uncommon. SBO may accelerate onset of spinal and neck pains [24,28]. Its discovery will answer the question of why premature degenerative disc disease in a young patient (Tables 1 and 2).

In terms of limitation, the present study was not population based and therefore could not provide any estimates of the incidence of spina bifida occulta in our region. Further studies with larger samples are needed to have a better insight into this disease in Uyo in lieu of hospital-based epidemiological study (Figures 1 and 2).

Table 1: Showing age distribution of studied population.

Age range	Males	Females	Total
0-9	0	0	0
10-19	0	4	4
20-29	10	8	18
30-39	22	6	28
40-49	10	10	20
50-59	14	24	38
60-69	12	4	16
70-79	8	0	8
80-89	6	2	8
Total	82	58	140

Table 2: Showing number of patients with cervical SBO localizations.

Site of lesions	No of patients
C1	0
C2	0
C3	0
C4	2
C5	6
C6	6
C7	12

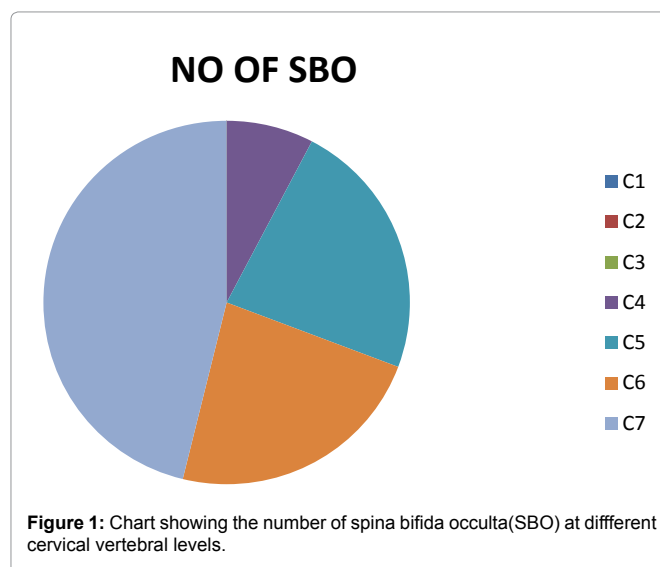


Figure 1: Chart showing the number of spina bifida occulta(SBO) at different cervical vertebral levels.

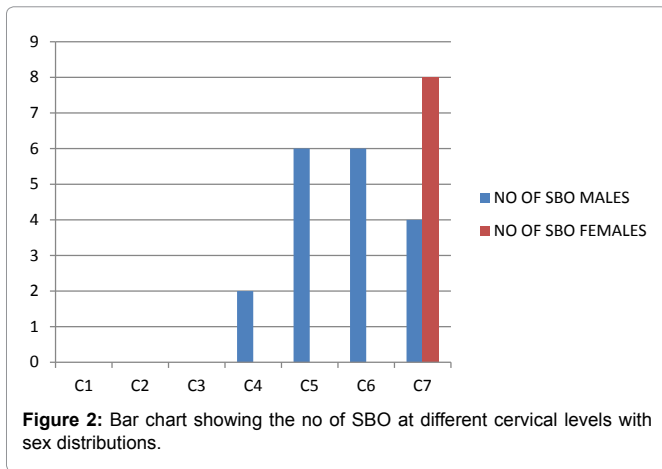


Figure 2: Bar chart showing the no of SBO at different cervical levels with sex distributions.

Conclusion

Incidence of cervical spina bifida occulta in University of Uyo teaching hospital is 12.86%. It has lower cervical vertebra preference, central localization restriction and female bias. The earliest age of radiographic discovery was in the middle age due to its asymptomatic nature. Spina bifida occulta of the cervical spine could be solitary or multiple. It is 100% solitary in females. The multiple types were more accompanied by cervical spondylosis.

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Author Affiliation

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¹Department of Radiology, Faculty of Clinical Sciences, University of Uyo, Nigeria

²Department of Radiation Medicine, Faculty of Health Sciences and Technology, College of Medicine, University of Nigeria, Enugu Campus, Nigeria

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