

Association of Lemon Sign of Fetal Skull With Spina Bifida

Lemon Sign of Fetal Skull With Spina Bifida

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ABSTRACT

Objective: To study Association of lemon sign of fetal skull with spina bifida

Study design: Observational study.

Place and Duration of Study: This study was conducted at Allama Iqbal Memorial Teaching Hospital Sialkot during Jan 2018 to Jan 2020.

Materials and methods; Fifty cases of Association of lemon sign of fetal skull with spina bifida, were included in this study. Ultrasonography was conducted for all the fifty cases showing lemon sign of fetal skull to study the association of lemon sign with the spina bifida. Written informed consent of the parents was taken before collection of data. Permission of ethical committee was also considered for collection of data and publishing in medical journal.

Results; Among fifty fetuses showing lemon sign of skull, forty nine showed spina bifida. Only one fetus demonstrating typical lemon shaped skull showed no evidence of spina bifida or associated abnormality. All the fetuses demonstrating spina bifida also shown associated features suggestive of Chiari malformations.

Conclusion: Lemon sign shows a strong association with spina bifida and spina bifida is strongly associated with Arnold Chiari malformation.

Key Words: Lemon sign, Skull, Spina bifida, Chiari Malformation

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INTRODUCTION

Spina bifida is a neural tube defect. It results from incomplete or aberrant fusion of posterior bony arches, anywhere along the spine¹. Earlier in 1970-80,s maternal serum alpha feto protein level was the main tool for screening the spina bifida by amniocentesis at/around 16 weeks². However invasive methods nearly replaced with development of better quality ultrasound machines and techniques. This better detection rate of spina bifida mainly attributed to the detection of abnormal cranial contour ,described as lemon sign^{3,4,5}. This sign depicts the abnormal fetal skull shaped due to symmetrical flattening /scalloping of bilateral frontal bones, alleviating the normal convex contour of bilateral frontal bones, giving lemon shaped architecture to skull^{4,6,7}. Many authors have shown the relation of lemon sign with spina bifida^{8,9}.

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In many cases, lemon sign is more conspicuous than spina bifida itself³, so after finding lemon shaped skull, a careful search can be made to visualize the spinal defect. Detection of spina bifida at an earlier gestation would be beneficial. Parents prefer to be informed as early as possible in cases of significant abnormalities. If ultimate choice of the parents is to terminate pregnancy, It would also be safer and easier to performed at earlier gestation. Early diagnosis of spina bifida also allows time for detailed counselling and assessment, if in-utero closure is being considered. It is found that fetal surgery to close spina bifida between 19 to 26 weeks has better out come as compared to postnatal repair¹⁰.

The present study is aimed to determine the validity of lemon sign in diagnosing the spina bifida. Result with high association of lemon sign and spina bifida will help to prevent missing of this anomaly in early stage of gestation, so that timely intervention in the form of In utero corrective procedures or therapeutic abortion can be planned.

MATERIALS AND METHODS

Fifty cases of Association of lemon sign of fetal skull with spina bifida, were included in this study. Ultrasonography was conducted for all the fifty cases demonstrating lemon sign of fetal skull to study the association of lemon sign with spina bifida. Findings of all the cases were correlated and reconfirmed by physical examination of newborn after delivery or therapeutic abortion. Written informed consent of the parents was taken before collection of data. Permission

of ethical committee was also considered for collection of data and publishing in medical journal.

Inclusion Criteria: Only those cases included in study, showing lemon shaped fetal skull on ultrasound.

Exclusion criteria: All those cases showing no evidence of lemon fetal shaped skull on ultrasound excluded from the study.

RESULTS

Among fifty cases showing lemon sign, forty nine cases showed evidence of spina bifida. Only one fetus showing lemon sign of skull showed no evidence of spina bifida as shown in Tab: 1.

Table No.1: Spina bifida in fetuses showing lemon sign.

Number of fetuses with lemon sign	Detection of Spina bifida	Percentage of detection
49	Detected	98%
1	Not detected	2%

Table No. 2: Location of spina bifida in fetuses showing lemon sign.

Location of Spina bifida	Number of fetuses	Location percentage
Thoracolumbar	28	57.2%
Lumbar	12	24.5%
Sacral	8	16.3%
Suboccipital/High cervical	1	2%

Table No. 3: Association of spina bifida with Chiari malformation.

Fetuses with Chiari malformation	Type of chiari malformation	Percentage
48	Type II	97.9%
1	Type III	2.1%

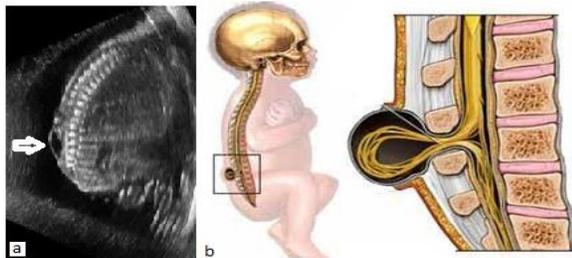


Figure No.1: Sagittal sonographic image showing meningocele, marked by arrow (a), illustration showing meningocele (b).

Distribution of location of spina bifida is shown in the table 2. All the cases showing spina bifida also showed associated features with variable severity like, small infra tentorial fossa, hydrocephalus , effacement of cisterna magna, banana shaped anteriorly concave cerebellum with cerebellar tonsillar herniation, suggestive

of Chiari malformation. Forty eight of these fetuses fall in category of Chiari II malformation and one of the fetus showing spina bifida with above mentioned associated features fall in Chiari III malformation, as shown in table 3.

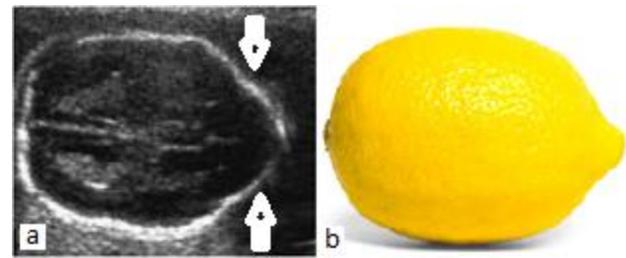


Figure No.2: Inward scalloping of frontal bones of fetal skull marked by arrows (a) ,illustration of lemon (b).

DISCUSSION

Neural tube malformations are the 2nd most common serious congenital defects after congenital heart anomalies¹¹. Worldwide prevalence of neural tube defect is 1-2 per 10,000 neonates¹². Spina bifida is a form of neural tube defect, that occurs due to the failure of fusion of neural tube by 28days of gestation, causing an opening of vertebra any where along the dorsal aspect of spine¹³. Spina bifida mainly classified into two groups: closed spina bifida (Spina bifida occulta) and Open spina bifida (Spina bifida aperta)¹⁴. Spina bifida occulta is a spinal malformations with intact overlying skin. Spina bifida occulta is rarely detected at early stage of gestation¹¹. It will not be discussed further in the present description. Spina bifida aperta involves exposure of the spinal cord and meninges with no intact overlying skin. Spina bifida aperta comprises of Myelocoele, meningocele and myeloschisis. A meningeal sac containing cerebrospinal fluid that protrudes out through the bony spinal defect is known meningocele, if neuronal elements like spinal cord and nerve roots also protrude into the sac, then it is called as meningocele, as shown in figure 1. In case of myeloschisis neural tissue flush with the surrounding skin¹³. The most common location of spina bifida is lumbosacral region, but flat or small lesions can be a real challenge to find out at any level¹⁵.

Spina bifida has a strong association with lemon sign of skull. The lemon sign represents the shape of the skull at ultrasound with medial displacement of bilateral frontal bones leading to inwards scalloping of frontal bones abolishing their normal convex contour. This gives a lemon like configuration to the fetal skull^{6,16}, shown in figure:2. To properly image this sign an axial image of fetal cranium is taken by ultrasound at the level of ventricles⁶.

Exact pathogenesis of this abnormal fetal cranial contour is unknown. However, it is hypothesized that in case of spina bifida, there is a decrease in intraspinal

pressure that causes the inferior displacement of hindbrain. It leads to decrease in intracranial pressure, which indirectly reflects as this abnormality of cranial contour, by inward scalloping of bilateral frontal bones. However with the progression of pregnancy the lemon sign disappears. The proposed reason behind this is that with advancement of gestation frontal bones become stronger enough to cope the stress of decreased pressure without deforming of the contour. The other described reason is that, majority of spina bifida cases also show progressing hydrocephalus. It causes increased intracranial pressure leading to reversal of inward scalloping of bilateral frontal bones to outward convex contour^{17,18}. However, this theory fails to explain the lemon sign in fetuses with normal posterior fossa. Therefore other opinion described by Furness and colleagues is that lemon like appearance of the fetal skull in cases of spina bifida might be result of mesenchymal dysplasia of cranium due to primary skeletal developmental disorder⁶.

Spina bifida also shows other associated abnormalities like hydrocephalus, obliteration/effacement of cisterna magna, abnormal anterior concavity or rolling of cerebellum around the brainstem (known as banana sign), most likely due to partial cerebellar tonsillar herniation through the foramen magnum, collectively these abnormalities described as Chiari malformation. With the continuous growth of the fetus, tethered cord at the level of spina bifida causes caudal traction of hindbrain, It might have a role in development of Chiari malformation^{3,19}. Prevalence of Chiari malformation is 0.1 to 0.5%²⁰.

Many investigators have described utility of lemon signs in detection of spina bifida. In 1986 Nicolaidis and coworkers in their retrospective study of 70 patients of open spina bifida, first of all described the term lemon sign²¹. In another study, Nyberg et al described that in fetuses with spina bifida, lemon sign had a high sensitivity (93% [13 of 14]), specificity (99% [212 of 215]) and a positive predictive value (81% [13 of 16])^{6,18}. However, a small percentage of structurally normal fetuses may demonstrate the lemon sign, as described by Campbell and associates, Nyberg and coworkers, Van den and co workers 1.2%, 1.3% and 0.66% respectively.^{18,22,23,24}

Rationale of this study was to gather and analyse data, about utility of lemon sign in detection of spina bifida in early gestation. In the present study fifty fetuses showing lemon shaped fetal skull before 24 weeks of gestation noted. Among these, forty nine fetuses showed open spina bifida. Only one fetus showing typical lemon like configuration of skull showed no abnormality of spine. A careful search was made to evaluate other associated abnormalities in this fetus during antenatal scans and after delivery neonate was physically examined but no gross abnormality noted. Excluding the terminated fetuses, the rest of the fetuses

show no evidence of lemon sign on ultrasound during latter stages of pregnancy. Among forty nine cases of spina bifida, twenty eight showed spina bifida at lumbosacral region, twelve fetuses showed at lumbar region, eight fetuses showed at sacral region and one fetus showed meningocele at suboccipital/high cervical region. All the fetuses showing spina bifida also showed features suggestive of Chiari malformation that include small infra tentorial fossa, hydrocephalous, effacement of cisterna magna, banana shaped anteriorly concave cerebellum with cerebellar tonsillar herniation. Forty eight fetuses showed Chiari II malformation, whereas one fetus showed Chiari III malformation. Chiari type II malformation is associated with lumbosacral spina bifida. Chiari type III malformation is a rare anomaly, associated with low occipital/high cervical meningocele/ meningoencephalocele⁷. Our results are almost similar with the already published data on the same subject^{6,18,21}. However, Present study in addition to suggest, the usefulness of lemon sign in early detection of spina bifida, also described a strong association of open spina bifida with Arnold Chiari malformation.

Main ultrasound feature of spina bifida is the splaying of the posterior elements of spine. At many instances it is difficult to diagnose this lesion in early pregnancy, especially in cases, when the fetuses are not in optimal position. Lesions of lower lumbar or sacral spine can be particularly challenging to diagnose in early period of gestation³. In these scenarios abnormal cranial contour in the form of lemon sign, strongly suggests careful evaluation, to find out likely spinal deformity.

CONCLUSION

Current study showed that though lemon sign not specific but is highly sensitive in diagnosing the spina bifida particularly in early pregnancy, where chance of missing the lesion is high. We also concluded in this study that there is a strong association of spina bifida with the Chiari malformation. It further stresses the need of early diagnosing the spina bifida by utilizing the lemon sign of fetal skull. Early diagnosis of spina bifida could have potentially important significance, It would allow the parents to gain sufficient time and information for management, which may include pregnancy termination, expectant, or fetal surgery.

Author's Contribution:

Concept & Design of Study:	Shams Munir
Drafting:	Ayesha Shams, Nasreen Hamid
Data Analysis:	Kamran Hamid, Muhammad Sabir
Revisiting Critically:	Shams Munir, Ayesha Shams
Final Approval of version:	Shams Munir

Conflict of Interest: The study has no conflict of interest to declare by any author.

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